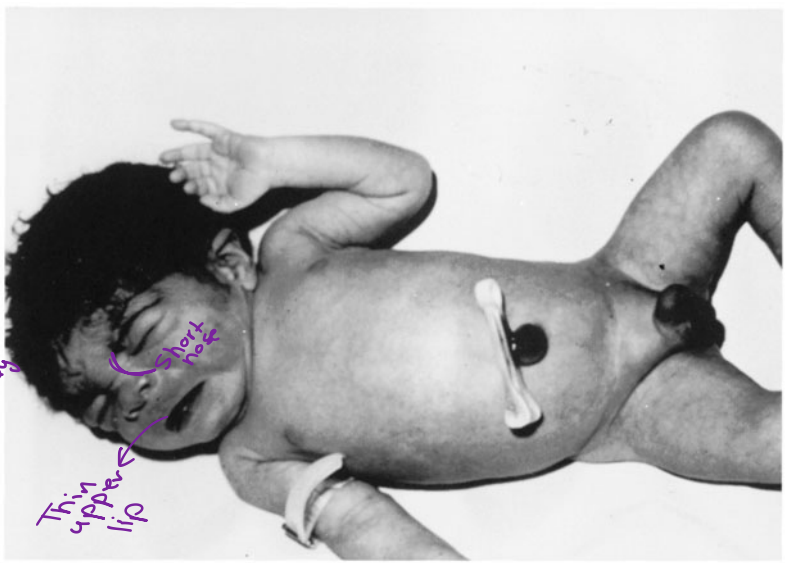
The Newborn Infant

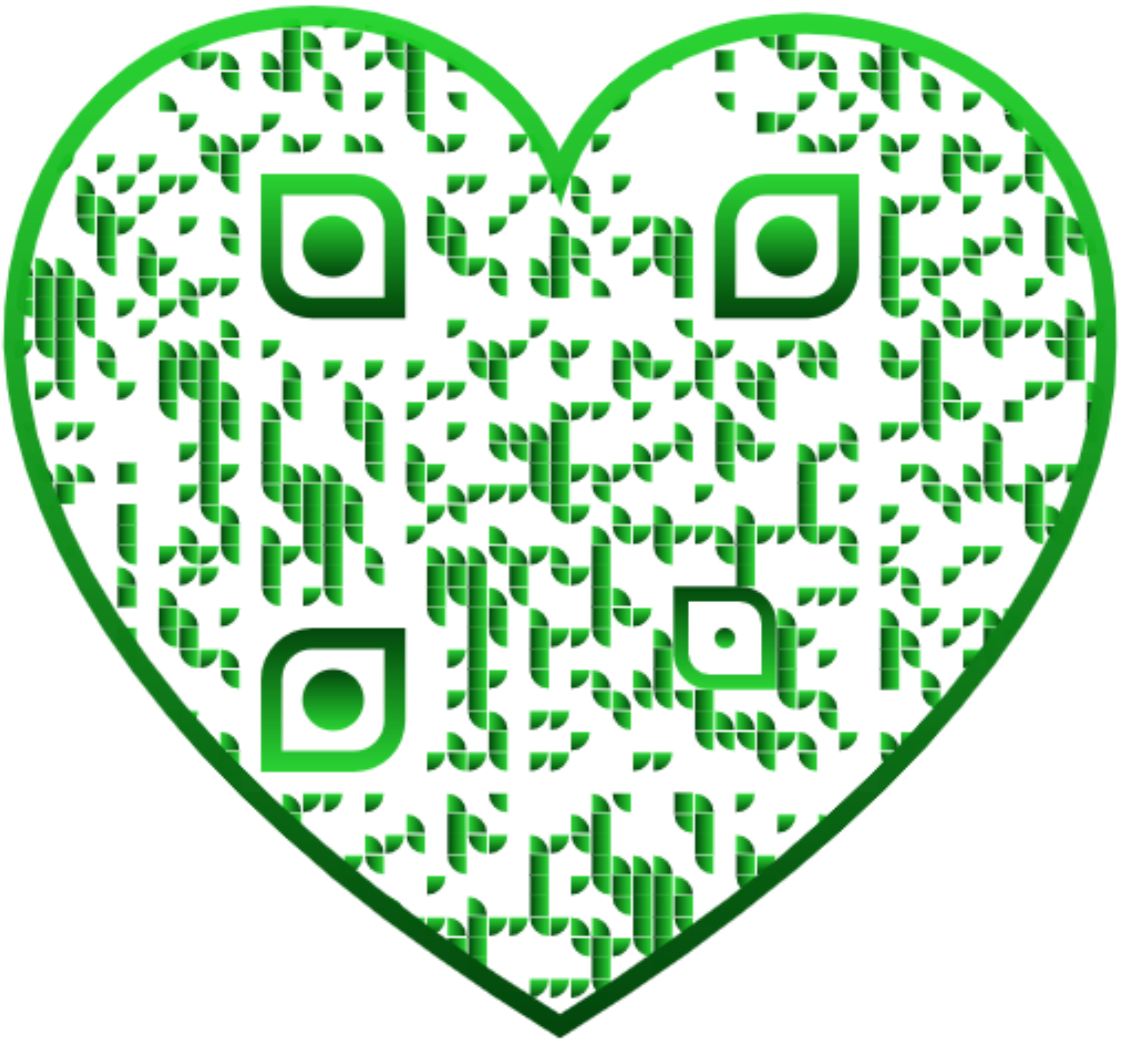
د. ايمان نبيل محمد
طب عام ج/ الكوفة

108. The term infant pictured below weighs 2200 g (4 lb, 14 oz). He is found to have a ventricular septal defect on cardiac evaluation. This infant appears to have features consistent with which of the following?



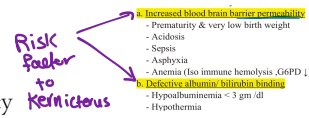
a. Perinatal phenytoin exposure
b. Trisomy 21
c. Alport syndrome
d. Fetal alcohol syndrome
e. Infant of diabetic mother

microcephaly, small palpebral fissures, short nose, smooth philtrum, thin upper lip, ptosis, microphthalmia, cleft lip and palate, and central nervous system abnormalities including mental retardation



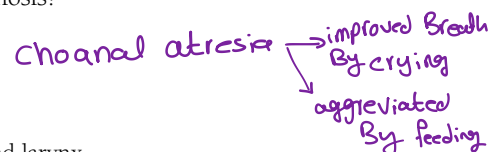
109. A newborn is noted to be quite jaundiced at 3 days of life. Laboratory data demonstrate his total bilirubin to be 17.8 mg/dL (direct bilirubin is 0.3 mg/dL). Which of the following factors is associated with an increased risk of neurologic damage in a jaundiced newborn?

a. Metabolic alkalosis
b. Increased attachment of bilirubin to binding sites caused by drugs such as sulfisoxazole
c. Hyperalbuminemia
d. Neonatal sepsis
e. Maternal ingestion of phenobarbital during pregnancy


Risk factor to Kernicterus
a. Increased blood brain barrier permeability
- Prematurity & very low birth weight
- Acidosis
- Sepsis
- Asphyxia
- Anemia (Iso immune hemolysis ,G6PD ↓)
b. Defective albumin/ bilirubin binding
- Hypoalbuminemia < 3 gm /dl
- Hypothermia

110. A 2-hour-old full-term newborn infant is noted by the nursing staff to be having episodes of cyanosis and apnea. Per nursery protocol they place an oxygen saturation monitor on him. When they attempted to feed him, his oxygen levels drop into the 60s. When he is stimulated and cries, his oxygen levels increase into the 90s. Which of the following is the most important next step to quickly establish the diagnosis?

a. Echocardiogram
b. Ventilation perfusion scan
c. Passage of catheter into nose
d. Hemoglobin electrophoresis
e. Bronchoscopic evaluation of palate and larynx



111. A mother calls you frantic because she has just been diagnosed with chicken pox. She delivered 7 days ago a term infant that appears to be eating, stooling, and urinating well. The child has been afebrile and seems to be doing well. Which of the following is the most appropriate step in management?

a. Isolate the infant from the mother.
b. Hospitalize the infant in the isolation ward.
c. Administer acyclovir to the infant.
d. Administer varicella-zoster immunoglobulin to the infant.
e. Advise the mother to continue regular well-baby care for the infant.

intervention محد يوصين عم الطفل وهو سليم فما داعي لأي

varicella-zoster immunoglobulin (VZIG) should be administered to the infant immediately after delivery if the mother had the onset of varicella within 5 days prior to delivery, and immediately upon diagnosis if her chicken pox started within 2 days after delivery.

If a normal full-term newborn is exposed to chicken pox 2 or more days postnatally, VZIG and isolation are not necessary because these babies appear to be at no greater risk for complications than older children. Acyclovir may be used in infants at risk for severe varicella, such as those infants exposed perinatally.

112. A mother wishes to breast-feed her newborn infant, but is worried about medical conditions that would prohibit her from doing so. You counsel her that of her listed conditions, which of the following is a contraindication to breast-feeding?

a. Upper respiratory tract infection
b. Cracked and bleeding nipples
c. Mastitis
d. Inverted nipples
e. HIV infection

113. A mother delivers a neonate with meconium staining and Apgar scores of 3 at 1 and 5 minutes of life. She had no prenatal care and the delivery was by emergency cesarean section for what the obstetricians report as "severe fetal bradycardia." Which of the following sequelae could be expected to develop in this intubated neonate with respiratory distress?

a. Sustained rise in pulmonary arterial pressure
b. Hyperactive bowel sounds
c. Microcephaly with micrognathia
d. Cataracts
e. Thrombocytosis

114. A 2-year-old boy is being followed for congenital cytomegalovirus (CMV) infection. He is deaf and developmentally delayed. The child's mother informs you that she has just become pregnant and is concerned that the new baby will be infected and may develop serious consequences. Which of the following is true?

a. The mother has antibodies to CMV that are passed to the fetus.
b. The mother's infection cannot become reactivated.
c. The likelihood that the new baby will become clinically ill is approximately 80%.
d. Termination of pregnancy is advised.
e. The new infant should be isolated from the older child.

Symptomatic congenital disease usually occurs when a mother has a primary CMV infection in the first trimester of pregnancy. Many of these babies die, and those who survive are severely affected. In the event of reactivation of CMV infection during pregnancy, maternal IgG, passed transplacentally, protects the infant from serious infection.

115. A full-term infant is born after a normal pregnancy; delivery, however, is complicated by marginal placental separation. At 12 hours of age, the child, although appearing to be in good health, passes a bloody meconium stool. For determining the cause of the bleeding, which of the following diagnostic procedures should be performed first?

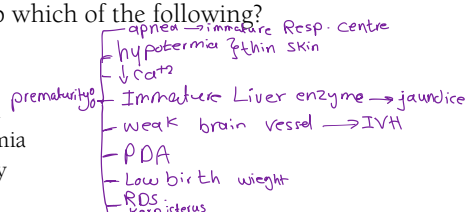
a. A barium enema
b. An Apt test
c. Gastric lavage with normal saline
d. An upper gastrointestinal series
e. A platelet count, prothrombin time, and partial thromboplastin time

116. As you are about to step out of a newly delivered mother's room, she mentions that she wants to breast-feed her healthy infant, but that her obstetrician was concerned about one of the medicines she was taking. Which of the woman's medicines, listed below, is clearly contraindicated in breast-feeding?

a. Ibuprofen as needed for pain or fever
b. Labetalol for her chronic hypertension
c. Amphetamines for her attention deficit disorder
d. Carbamazepine for her seizure disorder
e. Acyclovir for her HSV outbreak

117. A recovering premature infant who weighs 950 g (2 lb, 1 oz) is fed breast milk to provide 120 cal/kg/d. Over the ensuing weeks, the baby is most apt to develop which of the following?

a. Hypernatremia
b. Hypocalcemia
c. Blood in the stool
d. Hyperphosphatemia
e. Vitamin D toxicity



118. A primiparous woman whose blood type is O positive gives birth at term to an infant who has A-positive blood and a hematocrit of 55%. A total serum bilirubin level obtained at 36 hours of age is 12 mg/dL. Which of the following additional laboratory findings would be characteristic of ABO hemolytic disease in this infant?

a. A normal reticulocyte count
b. A positive direct Coombs test → in Rh / Abo incompatibility
c. Crescent-shaped red blood cells in the blood smear
d. Elevated hemoglobin
e. Petechiae

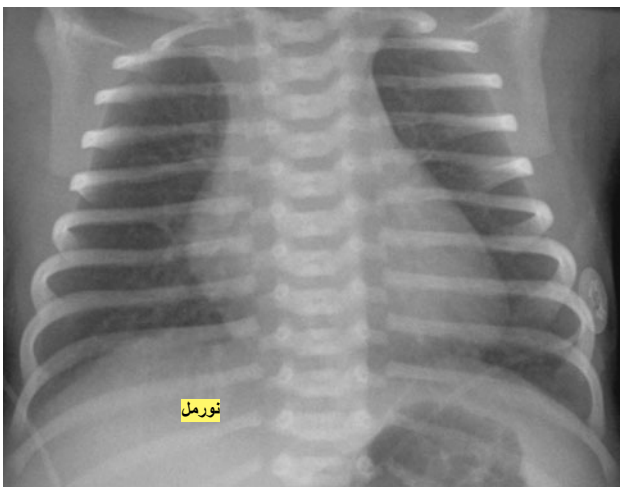
119. The nurse from the level 2 neonatal intensive care nursery calls you to evaluate a baby. The infant, born at 32 weeks' gestation, is now 1 week old and had been doing well on increasing nasogastric feedings. This afternoon, however, the nurse noted that the infant has vomited the last two feedings and seems less active. Your examination reveals a tense and distended abdomen with decreased bowel sounds. As you are evaluating the child, he has a grossly bloody stool. The plain film of his abdomen is shown. The next step in your management of this infant should include which of the following?

(Courtesy of Susan John, MD.)

a. Surgical consultation for an emergent exploratory laparotomy.
b. Continued feeding of the infant, as gastroenteritis is usually self-limited.
c. Stool culture to identify the etiology of the bloody diarrhea and an infectious diseases consultation.
d. Stopping feeds, beginning intravenous fluids, ordering serial abdominal films, and initiating systemic antibiotics.
e. Removal of nasogastric tube, placement of a transpyloric tube and, after confirmation via radiograph of tube positioning, switching feeds from nasogastric to nasoduodenal.

Management depends initially on the presence or absence of perforation;
if no evidence of free peritoneal air is found, the infant should be put on bowel rest with nasogastric decompression, and systemic antibiotics are initiated. Electrolytes and vital signs should be monitored closely, and serial abdominal films should be performed to evaluate for perforation.
If free air is identified on plain radiographs or if the infant clinically worsens with medical management, surgical consultation is required. An exploratory laparotomy is usually performed, and any necrotic intestinal tissue is removed. Occasionally, removal of necrotic gut will result in an infant without adequate intestinal surface area to absorb nutrition, a condition known as short bowel syndrome.

120. An infant weighing 1400 g (3 lb) is born at 32 weeks' gestation. Initial evaluation was benign, and the infant was transferred to the level 2 nursery for prematurity. The nurse there calls at 1 hour of life and reports the infant is tachypneic. Vital signs include a heart rate of 140 beats per minute, a respiratory rate of 80 breaths per minute, a temperature of 35°C (95°F), and a peripheral oxygen saturation of 98%. The lungs are clear with bilateral breath sounds and there is no murmur; the infant is in no distress. The child's chest radiograph is shown. Which of the following is the most appropriate next step in evaluating the infant?



(Courtesy of Susan John, MD.)

a. Obtain a complete blood count and differential.
b. Perform a lumbar puncture.
c. Administer intravenously 5cc of $D_{50}W$.
d. Place the infant under a warmer.
e. Administer supplemental oxygen.

121. Two new mothers are discussing their infants outside the neonatal intensive care unit. Both were born at 36 weeks' gestation. One infant weighs 2600 g (5 lb, 12 oz) while the other infant weighs 1600 g (3 lb, 8 oz). The mother of the second infant should be told that her child is more likely to have which of the following conditions?

a. Congenital malformations
b. Low hematocrit
c. Hyperglycemia
d. Surfactant deficiency
e. Rapid catch-up growth

The small-for-dates infants have a higher incidence of major congenital anomalies and are at increased risk for future growth retardation,

122. A 3-day-old infant, born at 32 weeks' gestation and weighing 1700 g (3 lb, 12 oz), has three episodes of apnea, each lasting 20 to 25 seconds and occurring after a feeding. During these episodes, the heart rate drops from 140 to 100 beats per minute, and the child remains motionless; between episodes, however, the child displays normal activity. Blood sugar is 50 mg/dL and serum calcium is normal. Which of the following is most likely true regarding the child's apneic periods?

a. They are due to an immature respiratory center.
b. They are a part of periodic breathing.
c. They are secondary to hypoglycemia.
d. They are manifestations of seizures.
e. They are evidence of underlying pulmonary disease.

123. You have an 11-day-old term infant in your office for a well-child visit. The mother notes that she received a letter that day from the state's Department of Health reporting that her child's newborn screen had come back abnormal, indicating possible galactosemia. Which of the following is the most appropriate management at this point?

a. Discontinue oral feeds and begin total parenteral nutrition.
b. Supplement her breast-feeding with a multivitamin.
c. Refer to endocrinology for evaluation.
d. Discontinue breast-feeding and initiate soy formula feedings.
e. Ultrasound of pancreas.

Galactosemia: Autosomal recessive disorder

Normally: Lactose $\xrightarrow[\text{enzyme}]{\text{Lactase}}$ Glucose + Galactose -1-Phosphate (Gal-1-P)

Gal-1-P $\xrightarrow[\text{Uridyle transferase}]{\text{Galactose 1 phosphate}}$ Glucose.

In galactosemia : absent Gal-1-P uridyl transferase→ accumulated Gal-1-P

leads to:
- Cataract (absent red reflex in newborn)
- Chronic active hepatitis, hepatomegaly
- Mental retardation

Treatment: lactose/galactose-free formula

124. The father of a 1-week-old infant comes to the office in a panic. He has just noticed on his child a right anterior shoulder mass that seems tender. The father is an osteosarcoma survivor and fears the child has the same malignancy. In reviewing the baby's discharge papers, you note the child was a term, appropriate-for-gestational-age vaginal delivery with a birth weight of 3200 g (7 lb, 1 oz). Apgar scores were 9 at 1 and 5 minutes. Your examination is significant for a large firm mass on the right clavicle; the rest of the examination is normal. Management of this problem should include which of the following?

a. Magnetic resonance imaging of the right shoulder
b. Reassurance and supportive care
c. A biopsy of the mass for culture and cytology
d. Referral to an orthopedic surgeon
e. Skin biopsy to test for osteogenesis imperfecta

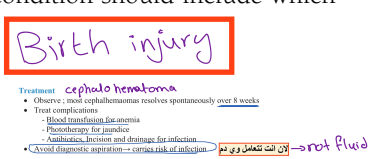
Clavicular fracture may happen in any delivery, although there is higher risk with large-for-gestational-age infants. Initial presentation of a fractured clavicle may include a pseudoparalysis, in which the infant refuses to move the ipsilateral arm, mimicking an Erb-Duchenne paralysis.

125. A 1-day-old infant who was born by a difficult forceps delivery is alert and active. She does not move her left arm spontaneously or during a Moro reflex. Rather, she prefers to maintain it internally rotated by her side with the forearm extended and pronated. The rest of her physical examination is normal. This clinical scenario most likely indicates which of the following?

a. Fracture of the left clavicle
b. Fracture of the left humerus
c. Left-sided Erb-Duchenne paralysis
d. Left-sided Klumpke paralysis
e. Spinal injury with left hemiparesis

126. You are examining a newborn infant in the well-baby nursery. The infant was the product of a benign pregnancy and vaginal delivery; he appears to be in no distress. Interestingly, your measurement of fronto-occipital head circumference is about 2 cm larger than the initial measurement done several hours before. Your examination otherwise is significant for tachycardia and a "squishy" feel to the entire scalp. You can elicit a fluid wave over the scalp. Management of this condition should include which of the following?

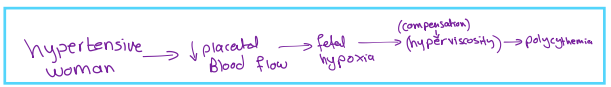
a. Transfer to the newborn ICU
b. Observation and parental reassurance
c. CT scan of the skull with bone windows
d. Surgical drainage
e. Elevation of the head of the crib


Birth injury
Treatment
cephalo hematoma
Observe ; most cephalhemaomas resolves spontaneously over 8 weeks
Treat complications
- Blood transfusion for anemia
- Phototherapy for jaundice
- Antibiotics, Incision and drainage for infection
Avoid diagnostic aspiration → carries risk of infection
لأن انت تتعامل وي دم
→ not fluid

A subgaleal hemorrhage will feel like a cephalohematoma that crosses the midline, but rapidly expands and can have cardiovascular complications. Careful monitoring is essential. Observation alone in this case would be appropriate, but should be accomplished in an ICU setting.

127. A 19-year-old primiparous woman develops toxemia in her last trimester of pregnancy and during the course of her labor is treated with magnesium sulfate. At 38 weeks' gestation, she delivers a 2100-g (4-lb, 10-oz) infant with Apgar scores of 1 at 1 minute and 5 at 5 minutes. Laboratory studies at 18 hours of age reveal a hematocrit of 79%, platelet count of 100,000/μL, glucose 41 mg/dL, magnesium 2.5 mEq/L, and calcium 8.7 mg/dL. Soon after, the infant has a generalized convulsion. Which of the following is the most likely cause of the infant's seizure?

a. Polycythemia
b. Hypoglycemia
c. Hypocalcemia
d. Hypermagnesemia
e. Thrombocytopenia



128. An infant who appears to be of normal size is noted to be lethargic and somewhat limp after birth. The mother is 28 years old, and this is her fourth delivery. The pregnancy was uncomplicated, with normal fetal monitoring prior to delivery. Labor was rapid, with local anesthesia and intravenous meperidine (Demerol) administered for maternal pain control. Which of the following therapeutic maneuvers is likely to improve this infant's condition most rapidly?

a. Intravenous infusion of 10% dextrose in water
b. Administration of naloxone (Narcan)
c. Administration of vitamin K
d. Measurement of electrolytes and magnesium levels
e. Neurologic consultation

129. At 43 weeks' gestation, a long, thin infant is delivered. The infant is apneic, limp, pale, and covered with "pea soup" amniotic fluid. Which of the following is the best first step in the resuscitation of this infant at delivery?

a. Intubation and suction of the trachea; provision of oxygen
b. Artificial ventilation with bag and mask
c. Chest compressions
d. Administration of 100% oxygen by mask
e. Catheterization of the umbilical vein

Meconium aspiration

130. The newborn pictured below was born at home and has puffy, tense eyelids; red conjunctivae; a copious amount of purulent ocular discharge; and chemosis 2 days after birth. Which of the following is the most likely diagnosis?

(Courtesy of Kathryn Musgrove, MD.)

a. Dacryocystitis
b. Chemical conjunctivitis
c. Pneumococcal ophthalmia
d. Gonococcal ophthalmia
e. Chlamydial conjunctivitis

131. After an uneventful labor and delivery, an infant is born at 32 weeks' gestation weighing 1500 g (3 lb, 5 oz). Respiratory difficulty develops immediately after birth and increases in intensity thereafter. At 6 hours of age, the child's respiratory rate is 60 breaths per minute. Examination reveals grunting, intercostal retraction, nasal flaring, and marked cyanosis in room air. Auscultation reveals poor air movement. Physiologic abnormalities compatible with these data include which of the following?

a. Decreased lung compliance, reduced lung volume, left-to-right shunt of blood
b. Decreased lung compliance, reduced lung volume, right-to-left shunt of blood
c. Decreased lung compliance, increased lung volume, left-to-right shunt of blood
d. Normal lung compliance, reduced lung volume, left-to-right shunt of blood
e. Normal lung compliance, increased lung volume, right-to-left shunt of blood

لان بضغر حجم الرئة فترجعه لهم القلب

132. A term infant is born to a known HIV-positive mother. She has been taking antiretroviral medications for the weeks prior to the delivery of her infant. Routine management of the healthy infant should include which of the following?

a. Admission to the neonatal intensive care unit for close cardiovascular monitoring
b. HIV ELISA on the infant to determine if congenital infection has occurred
c. A course of zidovudine for the infant
d. Chest radiographs to evaluate for congenital *Pneumocystis carinii*
e. Administration of IVIG to the baby to decrease the risk of perinatal HIV infection

133. Initial examination of a full-term infant weighing less than 2500 g (5 lb, 8 oz) shows edema over the dorsum of her hands and feet. Which of the following findings would support a diagnosis of Turner syndrome?

a. A liver palpable to 2 cm below the costal margin
b. Tremulous movements and ankle clonus
c. Redundant skin folds at the nape of the neck
d. A transient, longitudinal division of the body into a red half and a pale half
e. Softness of the parietal bones at the vertex

134. You have been recently named as the medical director of the normal newborn nursery in your community hospital and have been asked to write standardized admission orders for all pediatricians to follow. Which of the following vaccines will you include on these orders?

a. Hepatitis A vaccine
b. Hepatitis B vaccine
c. Combination diphtheria, tetanus, and acellular pertussis vaccine
d. Inactivated polio virus
e. Haemophilus influenza B vaccine

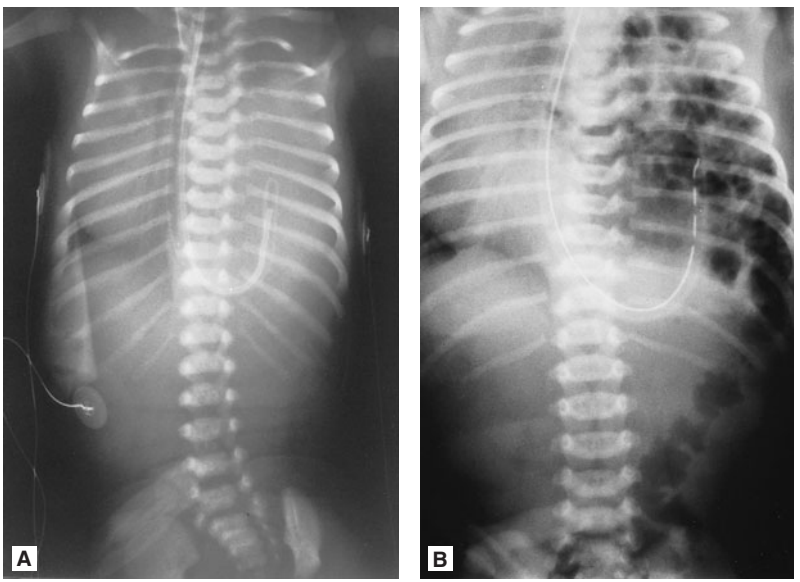
135. A 1-week-old black infant presents to you for the first time with a large, fairly well-defined, purple lesion over the buttocks bilaterally, as shown in the photograph. The lesion is not palpable, and it is not warm nor tender. The mother denies trauma and reports that the lesion has been present since birth. This otherwise well-appearing infant is growing and developing normally and appears normal upon physical examination. Which of the following is the most appropriate course of action in this infant?

(Courtesy of Adelaide Hebert, MD.)

Mongolian spot, a bluish-gray lesion located over the buttocks, lower back, and occasionally, the extensor surfaces of the extremities

a. Report the family to child protective services
b. Reassurance of the normalcy of the condition
c. Soft tissues films of the buttocks to identify calcifications
d. Administration of vitamin K
e. Measurement of bleeding time as well as factor VII and XI levels

136. A newborn infant develops respiratory distress immediately after birth. His abdomen is scaphoid. No breath sounds are heard on the left side of his chest, but they are audible on the right. Immediate intubation is successful with little or no improvement in clinical status. Emergency chest x-ray is shown (A) along with an x-ray 2 hours later (B). Which of the following is the most likely explanation for this infant's condition?



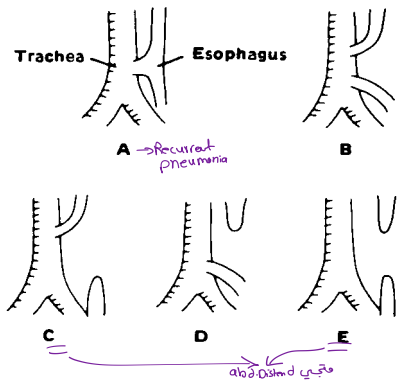
(Courtesy of Susan John, MD.)

a. Pneumonia
b. Cystic adenomatoid malformation
c. Diaphragmatic hernia
d. Choanal atresia
e. Pneumothorax

جراحه اطفال

137. Shortly after birth, an infant develops abdominal distention and begins to drool. When she is given her first feeding, it runs out the side of her mouth, and she coughs and chokes. Physical examination reveals tachypnea, intercostal retractions, and bilateral pulmonary rales. The esophageal anomaly that most commonly causes these signs and symptoms is illustrated by which of the following?

جراحه اطفال



a. Figure A
b. Figure B
c. Figure C
d. Figure D
e. Figure E

The anomaly illustrated by D is the most common; that of A can be diagnosed after repeated episodes of pneumonia. The anomalies in E and C are associated with all the same symptoms except abdominal distention, which cannot develop because air cannot enter the gastrointestinal tract. B and C are the least common; in these, the upper esophageal segment is connected directly to the trachea, and massive entry of fluid into the lungs occurs.

138. You are advised by the obstetrician that the mother of a baby she has delivered is a carrier of hepatitis B surface antigen (HBsAg-positive). Which of the following is the most appropriate action in managing this infant?

a. Screen the infant for HBsAg.
b. Isolate the infant with enteric precautions.
c. Screen the mother for hepatitis B "e" antigen (HBeAg).
d. Administer hepatitis B immune globulin and hepatitis B vaccine to the infant.
e. Do nothing because transplacentally acquired antibody will prevent infection in the infant.

139. You are called to a delivery of a term infant, about to be born via cesarean section to a mother with multiple medical problems, including a 1-month history of a seizure disorder, for which she takes phenytoin; rheumatic heart disease, for which she must take penicillin daily for life; hypertension, for which she takes propranolol; acid reflux, for which she takes aluminum hydroxide; and a deep venous thrombosis in her left calf diagnosed 2 days ago, for which she was started on a heparin infusion. The obstetrician is concerned about the possible effects of the mother's multiple medications on the newborn infant. Which of the following medications is most likely to cause harm in this newborn infant at delivery?

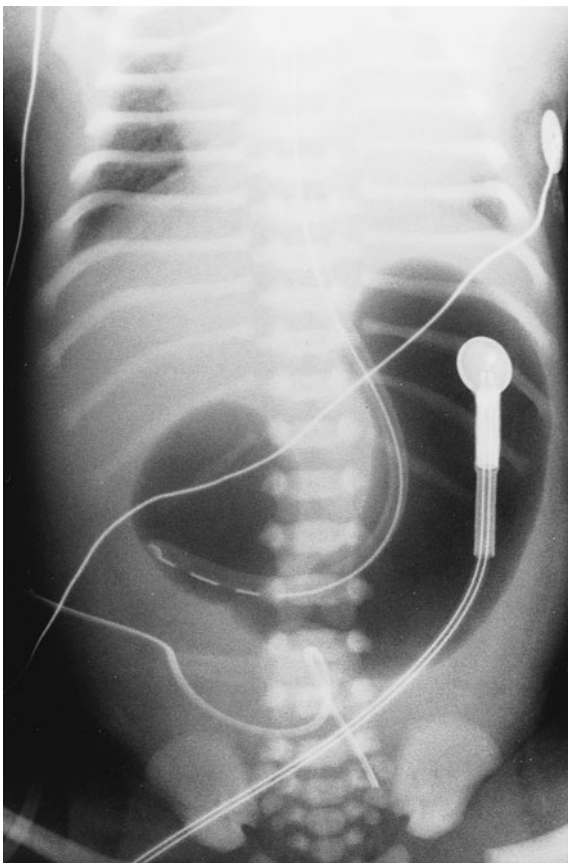
a. Propranolol
b. Penicillin
c. Aluminum hydroxide
d. Phenytoin
e. Heparin → Don't cross placenta

Propranolol, which may cause growth retardation when given throughout pregnancy, diminishes the ability of an asphyxiated infant to increase heart rate and cardiac output. It has also been associated with hypoglycemia and apnea.

140. Your older sister, her husband, their 2-day-old infant, and their pet Chihuahua arrive at your door. The parents of the child are concerned because the pediatrician noted the child was "yellow" and ordered some studies. They produce a wad of papers for you to review. Both the mother and baby have O-positive blood. The baby's direct serum bilirubin is 0.2 mg/dL, with a total serum bilirubin of 11.8 mg/dL. Urine bilirubin is positive. The infant's white blood cell count is 13,000/μL with a differential of 50% polymorphonuclear cells, 45% lymphocytes, and 5% monocytes. The hemoglobin is 17 g/dL, and the platelet count is 278,000/μL. Reticulocyte count is 1.5%. The peripheral smear does not show fragments or abnormal cell shapes. Which of the following is the most likely explanation for this infant's skin color?

a. Rh or ABO hemolytic disease
b. Physiologic jaundice 2nd Day
c. Sepsis
d. Congenital spherocytic anemia
e. Biliary atresia

141. At the time of delivery, a woman is noted to have a large volume of amniotic fluid. At 6 hours of age, her baby begins regurgitating small amounts of mucus and bile-stained fluid. Physical examination of the infant is normal, and an abdominal x-ray is obtained (see below). Which of the following is the most likely diagnosis of this infant's disorder?



(Courtesy of Susan John, MD.)

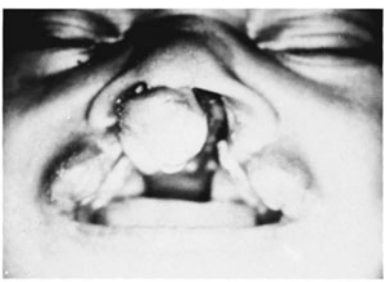
a. Gastric duplication
b. Pyloric stenosis
c. Esophageal atresia
d. Duodenal atresia
e. Midgut volvulus

142. The mother and father of a newborn come in for the 2-week checkup. The mother complains of "colic" and asks if she can switch to goat's milk instead of breast milk. Which of the following should be your main concern about using goat's milk instead of breast milk or cow's milk?

goat's Milk → Folate↓
cow Milk → iron↓

a. It has insufficient calories.
b. It has insufficient folate.
c. It has insufficient whey.
d. It has insufficient casein.
e. It has insufficient fat.

143. You see the newborn baby shown below for the first time in the nursery. You consult plastic and reconstructive surgeon as well as the hospital's speech therapist. Understandably, the parents have many questions. Which of the following statements is appropriate anticipatory guidance for this family?



a. Parenteral alimentation is recommended to prevent aspiration.
b. Surgical closure of the palatal defect should be done before 3 months of age.
c. Good anatomic closure will preclude the development of speech defects.
d. Recurrent otitis media and hearing loss are likely complications.
e. The chance that a sibling also would be affected is 1 in 1000.

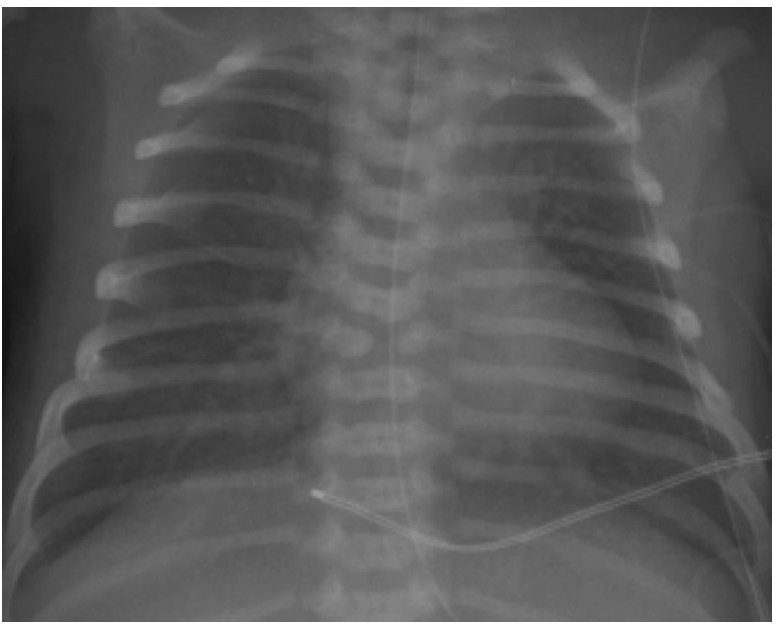
144. The mother of a 2-week-old infant reports that since birth, her infant sleeps most of the day; she has to awaken her every 4 hours to feed, and she will take only an ounce of formula at a time. She also is concerned that the infant has persistently hard, pellet-like stools. On your examination you find an infant with normal weight and length, but with an enlarged head. The heart rate is 75 beats per minute and the temperature is 35°C (95°F). The child is still jaundiced. You note large anterior and posterior fontanelles, a distended abdomen, and an umbilical hernia. This clinical presentation is likely a result of which of the following?

a. Congenital hypothyroidism
b. Congenital megacolon (Hirschsprung disease)
c. Sepsis
d. Infantile botulism
e. Normal development

145. A routine prenatal ultrasound reveals a male fetus with meningomyelocele. The 24-year-old primigravid mother is told the infant will require surgery shortly after birth. You counsel her about the etiology of this defect and the risk of further pregnancies being similarly affected, and state which of the following?

a. The hereditary pattern for this condition is autosomal recessive.
b. The prenatal diagnosis can be made by the detection of very low levels of alpha-fetoprotein in the amniotic fluid.
c. Subsequent pregnancies are not at increased risk compared to the general population.
d. Supplementation of maternal diet with folate leads to a decrease in incidence of this condition.
e. Neither environmental nor social factors have been shown to influence the incidence.

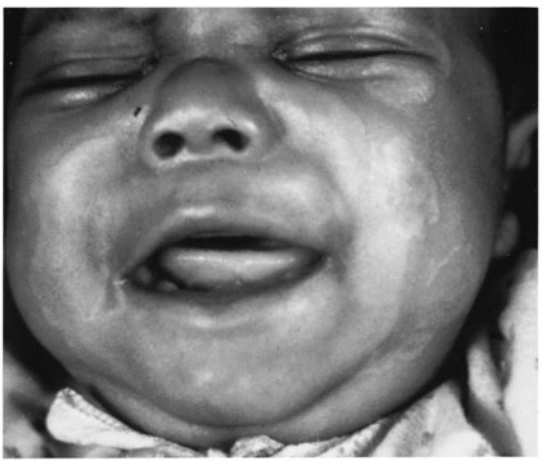
146. A term, 4200-g (9-lb, 4-oz) female infant is delivered via cesarean section because of cephalopelvic disproportion. The amniotic fluid was clear, and the infant cried almost immediately after birth. Within the first 15 minutes of life, however, the infant's respiratory rate increased to 80 breaths per minute, and she began to have intermittent grunting respirations. The infant was transferred to the level 2 nursery and was noted to have an oxygen saturation of 94%. The chest radiograph is shown. Which of the following is the most likely diagnosis?



(Courtesy of Susan John, MD.)

a. Diaphragmatic hernia
b. Meconium aspiration
c. Pneumonia
d. Idiopathic respiratory distress syndrome
e. Transient tachypnea of the newborn

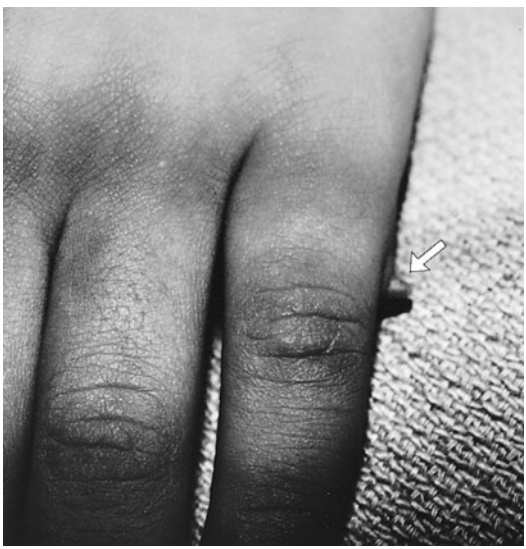
147. The infant in the following picture presents with hepatosplenomegaly, anemia, persistent rhinitis, and a maculopapular rash. Which of the following is the most likely diagnosis for this child?



(Courtesy of Adelaide Hebert, MD.)

a. Toxoplasmosis
b. Glycogen storage disease
c. Congenital hypothyroidism
d. Congenital syphilis
e. Cytomegalovirus disease

148. A well-appearing, 3200-g (7-lb, 1-oz) black infant is noted to have fifth finger (postaxial) polydactyly. The extra digit has no skeletal duplications and is attached to the rest of the hand by a threadlike soft tissue pedicle (see photograph). Appropriate treatment for this condition includes which of the following?



(Courtesy of Adelaide Hebert, MD.)

a. Chromosomal analysis
b. Excision of extra digit
c. Skeletal survey for other skeletal abnormalities
d. Echocardiogram
e. Renal ultrasound

149. An infant born at 35 weeks' gestation to a mother with no prenatal care is noted to be jittery and irritable, and is having difficulty feeding. You note coarse tremors on examination. The nurses report a high-pitched cry and note several episodes of diarrhea and emesis. You suspect which of the following?

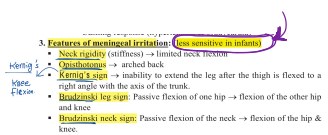
a. Fetal alcohol syndrome
b. Prenatal exposure to marijuana
c. Heroin withdrawal syndrome
d. Cocaine exposure *in utero*
e. Tobacco use by the mother

150. A previously healthy full-term infant has several episodes of duskiness and apnea during the second day of life. Diagnostic considerations should include which of the following?

a. Hemolytic anemia
b. Congenital heart disease
c. Idiopathic apnea
d. Harlequin syndrome
e. Hyperglycemia

151. The signs and symptoms of meningitis in an infant can be different than those in an adult. Which of the following signs and symptoms of meningitis is more helpful in an adult patient than in a 4-month-old?

a. Lethargy
b. Jaundice
c. Vomiting
d. Brudzinski sign
e. Hypothermia


3. Features of meningeal irritation: (less sensitive in infants)
Neck rigidity (stiffness) → limited neck flexion
Opisthotonus → arched back
Kernig's sign → inability to extend the leg after the thigh is flexed to a right angle with the axis of the trunk.
Brudzinski leg sign: Passive flexion of one hip → flexion of the other hip and knee
Brudzinski neck sign: Passive flexion of the neck → flexion of the hip & knee.
Kernig ↓
Knee flexion

meningitis, in a neonate is difficult because the signs and symptoms are subtle and nonspecific. They include lethargy; feeding problems including abdominal distention, vomiting, and diarrhea; temperature instability; respiratory distress or apnea; and jaundice. Nuchal rigidity and Kernig and Brudzinski signs are frequently not present in the neonate with meningitis.

152. A woman gives birth to twins at 38 weeks' gestation. The first twin weighs 2800 g (6 lb, 3 oz) and has a hematocrit of 70%; the second twin weighs 2100 g (4 lb, 10 oz) and has a hematocrit of 40%. Which of the following statements is correct?

a. The second twin is at risk for developing respiratory distress, cyanosis, and congestive heart failure.
b. The first twin is more likely to have hyperbilirubinemia and convulsions.
c. The second twin is at risk for renal vein thrombosis.
d. The second twin probably has hydramnios of the amniotic sac.
e. The second twin is likely to be pale, tachycardic, and hypotensive.

153. Parents bring a 5-day-old infant to your office. The mother is O negative and was Coombs positive at delivery. The term child weighed 3055 g (6 lb, 1 oz) at birth and had measured baseline hemoglobin of 16 g/dL and a total serum bilirubin of 3 mg/dL. He passed a black tarlike stool within the first 24 hours of life. He was discharged at 30 hours of life with a stable axillary temperature of 36.5°C (97.7°F). Today the infant's weight is 3000 g, his axillary temperature is 35°C (95°F), and he is jaundiced to the chest. Parents report frequent yellow, seedy stool. You redraw labs and find his hemoglobin is now 14 g/dL, and his total serum bilirubin is 13 mg/dL. The change in which of the following parameters is of most concern?

a. Hemoglobin
b. Temperature
c. Body weight
d. Bilirubin
e. Stool

temperature instability in a term infant is frequently a sign of serious infection.

154. You are called to a delivery of a woman with no prenatal care; she is in active labor but has no history of amniotic rupture. The biophysical profile done in the emergency center revealed severe oligohydramnios. When you get this infant to the nursery, you should carefully evaluate him for which of the following?

a. Anencephaly
b. Trisomy 18
c. Renal agenesis
d. Duodenal atresia
e. Tracheoesophageal fistula

155. A newborn infant becomes markedly jaundiced on the second day of life, and a faint petechial eruption, first noted at birth, is now a generalized purpuric rash. Hematologic studies for hemolytic diseases are negative. Acute management should include which of the following?

a. Liver ultrasound
b. Isolation of the infant from pregnant hospital personnel
c. Urine drug screen on the infant
d. Discharge with an early follow-up visit in 2 days to recheck bilirubin
e. Thyroid hormone assay

the patient described most likely has a congenital or acquired infection requiring immediate diagnosis

Questions 156 to 159

For each clinical scenario below, select the most likely diagnosis. Each lettered option may be used once, more than once, or not at all.

a. Congenital toxoplasmosis
b. Congenital syphilis
c. Congenital rubella
d. Congenital cytomegalovirus
e. Congenital herpes simplex virus

C **156.** A newborn has bilateral cataracts and microphthalmia, intrauterine growth retardation, hemorrhagic skin lesions scattered throughout the body, and a harsh systolic murmur heard at the left sternal border and radiating to the lung fields.

E **157.** A week-old infant presents with fever and focal seizure.

a **158.** A newborn has hydrocephalus, chorioretinitis, intracranial calcifications, and anemia.

d **159.** A newborn has microcephaly, intracranial calcifications, hepatosplenomegaly, and marked hyperbilirubinemia and thrombocytopenia.

Questions 160 to 163

For each clinical scenario below, select the most likely diagnosis. Each lettered option may be used once, more than once, or not at all.

a. Bronchopulmonary dysplasia
b. Respiratory distress syndrome (hyaline membrane disease)
c. Pulmonary interstitial emphysema
d. Bronchiolitis
e. Primary pulmonary hypoplasia
f. Pneumothorax
g. Asthma
h. Meconium aspiration
i. Phrenic nerve paralysis
j. Bacterial pneumonia

160. A large-for-gestation-age term infant is delivered via scheduled cesarean section develops, at 15 minutes of age, tachypnea, grunting, flaring, and retractions. The child does not move his left arm well, but you find no clavicular fracture. A chest radiograph shows the left diaphragm to be markedly higher than the right.

161. A postterm infant is born at home after a prolonged and difficult labor. The maternal grandmother brings the infant to the hospital at 1 hour of life because of fast breathing. Grandmother notes that the child seemed well for a while, but then developed increased work of breathing. Physical examination reveals an infant in moderate respiratory distress with diminished breath sounds on the left. Chest radiograph reveals the heart to be pushed to the right side and loss of lung markings in the left lung field.

162. An infant of uncertain but seemingly term dates is born via emergent cesarean section for nonreassuring heart tones; the obstetrician has noted little or no amniotic fluid. The infant is small, has abnormally shaped limbs, and an unusual facies. The child has immediate respiratory distress. A chest radiograph reveals a poorly developed chest with little lung tissue.

163. A preterm infant is now 7 weeks old. She was intubated for 2 weeks and was weaned off oxygen at 3 weeks of age. You are about to leave your office for Thanksgiving holiday when the emergency room calls to tell you she has new hypoxia, respiratory distress, wheezes, and runny nose. A chest radiograph reveals patchy infiltrates and hyperexpansion in both lung fields. The newborn's 2-year-old sibling has an upper respiratory infection.

Questions 164 to 167

Blood samples of a 3-day-old full-term infant are sent for screening to identify diseases that would have serious, permanent consequences without prompt and appropriate treatment. Select the most appropriate treatment for each disease below. Each lettered option may be used once, more than once, or not at all.

a. Special infant formula
b. Hormone therapy
c. Vitamin therapy
d. Antibiotic prophylaxis
e. Sunlight

The answers are 164-a, 165-a, 166-c, 167-b.

164. Galactosemia a → Galactose Free formula, avoid breast feeding

165. Phenylketonuria a

166. Biotinidase deficiency c

167. Hypothyroidism b

Questions 168 to 171

For each of the following descriptions of a patient with a congenital anomaly, select the major abnormality with which it is most likely to be associated. Each lettered option may be used once, more than once, or not at all.

a. Deafness
b. Seizures
c. Wilms tumor
d. Congestive heart failure
e. Optic glioma

The answers are 168-c, 169-a, 170-b, 171-b.

168. A child's left arm and leg seem bigger than those on the right. In addition, the child has aniridia. None of the family members have aniridia or hemihypertrophy, nor do they know of anyone else in the family with these conditions.

169. An infant has fusion of the eyebrows, heterochromic irises, a broad nasal root with lateral displacement of the medial canthi, and a white forelock.

170. An infant presents with a large, flat vascular malformation over the left face and scalp. The mother notes that her other child was born with a capillary hemangioma on his arm and asks if this is the same thing. You explain that this vascular malformation is different, and that you will want to monitor him for another condition.

171. A new mother points out several hypopigmented oval macules over the child's trunk and extremities. She notes that these have been present since birth. She is concerned because she had a brother with the same spots whom she thinks had "growths" in his brain.

Questions 172 to 174

For each description of head injury that follows, select the major abnormality with which it is most likely to be associated. Each lettered option may be used once, more than once, or not at all.

a. Intraventricular hemorrhage
b. Caput succedaneum
c. Subdural hemorrhage
d. Subarachnoid hemorrhage
e. Subgaleal hemorrhage

The answers are 172-e, 173-c, 174-a.

172. A 1-day-old infant has a fronto-occipital head circumference that is 2 cm larger than the initial measurement done several hours before, the scalp has a "squishy" feel to it, and the infant has developed tachycardia.

173. A 6-month-old comatose infant has multiple broken bones in various stages of healing, a bulging anterior fontanelle, and retinal hemorrhages.

174. Previous premature infant born at 27 weeks' gestation and now 6 months of age presents with macrocephaly and hydrocephalus on ultrasonogram.

تم بحمدالله وتوفيقه

د. ايمان نبيل محمد
طب عام ج/ الكوفة

The Newborn Infant

Answers

108. The answer is d. *(Hay et al, pp 17-18, 99-100, 474, 1031, 1045-1046. Kliegman et al, pp 507-509, 692, 780-785, 2172-2173. McMillan et al, pp 338, 423-427, 1875-1878, 2632, 2635. Rudolph et al, pp 124-126, 527, 732-734, 775, 776, 1699-1700.)* Fetal alcohol syndrome is a preventable cause of birth defects. Prenatal exposure to ethanol is the cause. Findings include small-for-gestation birth, microcephaly, small palpebral fissures, short nose, smooth philtrum, thin upper lip, ptosis, microphthalmia, cleft lip and palate, and central nervous system abnormalities including mental retardation (average IQ = 67).

Common findings with trisomy 21 include protruding tongue, Brushfield spots, redundant neck skin, mental retardation, brachycephaly, upslanting palpebral fissures, epicanthal folds, flat face, small ears, cardiac abnormalities (especially ventricular septal defect or endocardial Cushing defect), palmar creases, and clinodactyly of the fifth digit.

Dilantin exposure causes midface hypoplasia, low nasal bridge, ocular hypertelorism, and accentuated Cupid's bow of the upper lip. Other features include cleft lip and palate, growth retardation, mental deficiency, distal phalangeal hyperplasia, cardiovascular anomalies, and skeletal defects.

Alport syndrome is the most common of the hereditary nephritis conditions, frequently leading to end-stage renal disease. In 85% of patients with Alport syndrome, an X-linked dominant form of inheritance is found; about 15% are autosomal recessive. All cause hematuria and progressive nephritis. Other findings include deafness and ocular defects.

Infants of diabetic mothers have an increased chance of congenital heart disease, caudal regression syndrome, and a small left colon. Besides, they are large-for-gestation age and have a number of biochemical abnormalities such as hypoglycemia or hypocalcemia.

109. The answer is d. *(Hay et al, pp 11-17. Kliegman et al, pp 756-766. McMillan et al, pp 235-245. Rudolph et al, pp 164-169.)* Significant unconjugated serum bilirubin levels in full-term newborn infants can lead to diffusion of bilirubin into brain tissue and to neurologic damage. Sulfisoxazole and other drugs compete with bilirubin for binding sites on albumin;

therefore, the presence of these drugs can cause dislocation, not increased affinity, of bilirubin to tissues. Metabolic acidosis also reduces binding of bilirubin, and neonatal sepsis interrupts the blood–brain barrier, thus allowing diffusion of bilirubin into the brain. Administration of phenobarbital has been used to induce glucuronyl transferase in newborn infants and can reduce, rather than exacerbate, neonatal jaundice. Other factors that reduce the amount of unconjugated bilirubin bound to albumin (and therefore cause an increase in free unconjugated bilirubin) include hypoalbuminemia and certain compounds (eg, nonesterified fatty acids, which are elevated during cold stress) that compete with bilirubin for albumin-binding sites.

110. The answer is c. *(Hay et al, p 479. Kliegman et al, p 1743. McMillan et al, p 197. Rudolph et al, p 1260.)* It is important to make the diagnosis of choanal atresia quickly because it responds to treatment but can be lethal if unrecognized and untreated. Most neonates are obligate nose breathers and cannot breathe adequately through their mouths. Infants with choanal atresia have increased breathing difficulty during feeding and sleeping and improved respirations when crying. A variety of temporizing measures to maintain an open airway have been used, including oropharyngeal airways, positioning, tongue fixation, and endotracheal intubation, but surgical correction with placement of nasal tubes is most effective. The diagnosis can be made by failure to pass a catheter through the nose to the pharynx or by checking for fog developing on a cold metal instrument placed under each nares. Bronchoscopy would help diagnose lower airway anomalies. A ventilation-perfusion scan would be the appropriate examination if you were concerned about mismatch (eg, that associated with pulmonary embolism). A newborn with a hemoglobinopathy such as sickle cell or thalassemia would not present as this infant did, so an electrophoresis would not be helpful. An echocardiogram would be useful if you suspected congenital cyanotic heart disease. The lack of a murmur in a newborn does not rule out pathology; this would be a reasonable next step if the catheter passed through both nares without difficulty.

111. The answer is e. *(Hay et al, pp 1117-1119. Kliegman et al, pp 1367-1368. McMillan et al, pp 520-522. Rudolph et al, pp 1043, 1224.)* Per CDC recommendations, varicella-zoster immunoglobulin (VZIG) should be administered to the infant immediately after delivery if the mother had the onset of varicella within 5 days prior to delivery, and immediately upon diagnosis if her chicken pox started within 2 days after delivery. If untreated, about half of these infants will develop serious varicella as early as 1 day of age.

If a normal full-term newborn is exposed to chicken pox 2 or more days postnatally, VZIG and isolation are not necessary because these babies appear to be at no greater risk for complications than older children. Acyclovir may be used in infants at risk for severe varicella, such as those infants exposed perinatally.

112. The answer is e. *(Hay et al, pp 299-301. Kliegman et al, pp 215-216, 218. McMillan et al, p 116. Rudolph et al, p 104.)* There are few contraindications to breast-feeding. Active pulmonary tuberculosis and HIV are two examples of infectious contraindications in developed countries, as well as malaria, typhoid fever, and septicemia. In underdeveloped countries, the risk of infectious diarrhea due to use of contaminated water to mix formula or the unavailability of formula can preclude this recommendation. All medications taken by the mother will be secreted in breast milk, but usually not in amounts significant enough to affect the infant. Mothers taking antineoplastic agents should not breast-feed. Mothers with mastitis can continue to breast-feed; frequent feedings may help the condition by preventing engorgement. Mothers with mild viral illness may also continue to breast-feed. Cracked or bleeding nipples may make breast-feeding uncomfortable, but are not contraindications. Inverted nipples usually can be remedied, and only rarely prohibit breast-feeding.

113. The answer is a. *(Hay et al, pp 551-552. Kliegman et al, p 1727. McMillan et al, pp 315-319. Rudolph et al, pp 98-99, 188.)* The low Apgar scores, meconium staining, and ensuing respiratory distress suggest that asphyxia has occurred. During a period of asphyxia, the resulting hypoxemia, acidosis, and poor perfusion can damage a neonate's brain, heart, kidney, liver, and lungs. The resulting clinical abnormalities include cerebral edema, irritability, seizures, cardiomegaly, heart failure, renal failure, poor liver function, disseminated intravascular coagulopathy, and respiratory distress syndrome. There can be excessively high pulmonary arterial pressure at the same time systemic blood pressure begins to fall, resulting in a persistent right-to-left shunt across a patent ductus arteriosus or foramen ovale.

114. The answer is a. *(Hay et al, pp 1120-1123. Kliegman et al, pp 1377-1379. McMillan et al, pp 511-516. Rudolph et al, pp 1031-1035.)* Cytomegalovirus infection is the most common cause of congenital infection, occurring in 0.2% to 2.4% of all live births. Cytomegalic inclusion disease is a constellation

of findings including hepatomegaly, splenomegaly, jaundice, petechiae, purpura, and microcephaly. In the United States, 20% to 90% of women of childbearing age have serologic evidence of a past infection with CMV. Symptomatic congenital disease usually occurs when a mother has a primary CMV infection in the first trimester of pregnancy. Many of these babies die, and those who survive are severely affected. In the event of reactivation of CMV infection during pregnancy, maternal IgG, passed transplacentally, protects the infant from serious infection. Although most infants infected during this secondary maternal infection are asymptomatic, about 10% of them eventually manifest hearing and neurologic problems. Some recommend keeping a child with congenital CMV infection away from susceptible pregnant (or about to become pregnant) women because CMV excretion can persist for months to years; at the very least, good hand-washing practices should be instituted. If infected shortly after birth, the younger sibling will probably be asymptomatic since he or she has maternal IgG in the circulation. CMV is primarily an occult infection. Of toddlers in day-care centers, 20% to 80% acquire CMV and shed it in saliva and urine for years. Diagnosis is made with isolation of the virus from urine, saliva, or other secretions, although several rapid tests also are available.

115. The answer is b. *(Hay et al, p 631. Kliegman et al, p 774. McMillan et al, pp 1928-1929. Rudolph et al, p 1372.)* Hematemesis and melena are not uncommon in the neonatal period, especially if gross placental bleeding has occurred at the time of delivery. The diagnostic procedure that should be done first is the Apt test, which differentiates fetal from adult hemoglobin in a bloody specimen. The test is based upon the finding that fetal hemoglobin is alkali resistant, while adult hemoglobin will convert to hematin upon exposure to alkali. If the blood in an affected infant's gastric contents or stool is maternal in origin, further workup of the infant is obviated.

116. The answer is c. *(Hay et al, p 301. Kliegman et al, p 682. McMillan et al, p 198.)* Most medications are secreted to some extent in breast milk, and some lipid-soluble medications may be concentrated in breast milk. Although the list of contraindicated medications is short, caution should always be exercised when giving a medication to a breast-feeding woman. Medications that are clearly contraindicated include lithium, cyclosporin, antineoplastic agents, illicit drugs including cocaine and heroin, amphetamines, lithium, ergotamines, and bromocriptine (which suppresses lactation). Although some suggest that oral contraceptives may have a negative impact on milk

production, the association has not been proven conclusively. In general, antibiotics are safe, with only a few exceptions (such as tetracycline). While sedatives and narcotic pain medications are probably safe, the infant must be observed carefully for sedation. All of the medications listed in the question are considered safe, except for amphetamines.

117. The answer is b. *(Hay et al, pp 33-34. Kliegman et al, pp 706-707. McMillan et al, pp 180-181.)* It usually is impossible with any combination of parenteral and enteral nutrition to match what the infant would have accumulated *in utero*. The average, healthy, low-birth-weight infant of this size requires a daily intake of calcium of about 200 mg/kg. Breast milk has much less calcium (and phosphorus) than do commercial formulas. The breast milk can be directly supplemented with calcium, or it can be supplemented with commercial fortifiers. Alternatively, it can be mixed with formulas designed for the premature infant. Breast milk promotes gut maturation and prevents intestinal atrophy induced by lack of enteral feeding; however, it is likely to have insufficient calcium and phosphorus for catch-up growth.

118. The answer is b. *(Hay et al, p 13. Kliegman et al, p 772. McMillan et al, pp 242-243. Rudolph et al, pp 177, 1531-1532.)* If a mother is O positive and her baby is A positive, the baby has a chance of developing hemolytic disease. Hemolytic disease and jaundice caused by a major blood-group incompatibility are usually less severe than with Rh incompatibility. Although the hematocrit of affected infants usually is normal, elevation of the reticulocyte count and the presence of nucleated red blood cells and microspherocytes in the blood smear provide evidence of hemolysis. In comparison with hemolytic disease caused by Rh incompatibility, where it is usually strongly positive, major blood-group incompatibility is often associated with a direct Coombs test that is frequently weakly positive. Petechiae are usually associated with decreased number of platelets. Crescent-shaped (sickled) red blood cells are found with sickle-cell disease.

119. The answer is d. *(Hay et al, pp 38-39. Kliegman et al, pp 755-756. McMillan et al, pp 389-397. Rudolph et al, pp 140-143.)* The infant presented in the question has necrotizing enterocolitis (NEC), a potentially life-threatening disease of the neonate; the radiograph demonstrates distended loops of bowel with air in the bowel wall. NEC is more common in premature infants, but has been described in term infants as well. Although several organisms have

been isolated from NEC patients, no clear cause for this condition has been identified. Patients present with feeding intolerance and a distended abdomen; about a quarter have grossly bloody stool. Pneumatosis intestinalis is found on plain radiograph of the abdomen and is diagnostic for NEC in this age group. Management depends initially on the presence or absence of perforation; if no evidence of free peritoneal air is found, the infant should be put on bowel rest with nasogastric decompression, and systemic antibiotics are initiated. Electrolytes and vital signs should be monitored closely, and serial abdominal films should be performed to evaluate for perforation. If free air is identified on plain radiographs or if the infant clinically worsens with medical management, surgical consultation is required. An exploratory laparotomy is usually performed, and any necrotic intestinal tissue is removed. Occasionally, removal of necrotic gut will result in an infant without adequate intestinal surface area to absorb nutrition, a condition known as *short bowel syndrome*.

120. The answer is d. *(Hay et al, p 32. Kliegman et al, p 724. McMillan et al, p 259. Rudolph et al, pp 84-85, 123.)* The radiograph is normal. However, the vignette describes a cold infant. A room temperature of 24°C (approximately 75°F) provides a cold environment for newborn infants. Aside from the fact that these infants emerge from a warm, 37.6°C (99.5°F) intrauterine environment, at birth, infants (and especially preterm infants) are wet, have a relatively large surface area for their weight, and have little subcutaneous fat. Within minutes of delivery, the infants are likely to become pale or blue and their body temperatures will drop. In order to bring body temperature back to normal, they must increase their metabolic rate; ventilation, in turn, must increase proportionally to ensure an adequate oxygen supply. Because a preterm infant is likely to have respiratory problems and be unable to oxygenate adequately, lactate can accumulate and lead to a metabolic acidosis. Infants rarely shiver in response to a need to increase heat production. If the tachypnea persists after warming the infant, sepsis, pneumonia, and primary surfactant deficiency are all possible; thus, several of the alternative answers then may be appropriate.

121. The answer is a. *(Kliegman et al, p 703. McMillan et al, pp 274-181. Rudolph et al, pp 122, 2022-2023.)* Small-for-dates infants are subject to a different set of complications than preterm infants whose size is appropriate for gestational age. The small-for-dates infants have a higher incidence of major congenital anomalies and are at increased risk for future growth retardation, especially if length and head circumference as well as weight are small for

gestational age. Also more common are neonatal asphyxia and the meconium aspiration syndrome, which can lead to pneumothorax, pneumomediastinum, or pulmonary hemorrhage. These, rather than hyaline membrane disease, are the major pulmonary problems in small-for-gestational-age infants. Because neonatal symptomatic hypoglycemia is more commonly found in small-for-dates infants, careful blood glucose monitoring and early feeding are appropriate precautions. Normal or elevated hematocrit is also more common in these infants.

122. The answer is a. *(Hay et al, pp 34-35. Kliegman et al, pp 729-730. McMillan et al, pp 318-320. Rudolph et al, p 1934.)* Apneic episodes are characterized by an absence of respirations for more than 20 seconds and may be accompanied by bradycardia and cyanosis. A large number of conditions can cause apnea. Periods of apnea are generally thought to be secondary to an incompletely developed respiratory center, particularly when they are seen, as is common, associated with prematurity. Although seizures, hypoglycemia, and pulmonary disease accompanied by hypoxia can lead to apnea, these causes are less likely in the infant described, given that no unusual movements occur during the apneic spells, the blood sugar level is more than 40 mg/dL, and the child appears well between episodes. Other, less common, explanations for central apnea include congenital central hypoventilation syndrome (formerly known as Ondine's curse), Arnold-Chiari malformations, and congenital infections. Periodic breathing, a common pattern of respiration in low-birth-weight babies, is characterized by recurrent breathing pauses of 3 to 10 seconds.

123. The answer is d. *(Hay et al, pp 992-993. Kliegman et al, pp 609-610. McMillan et al, pp 2187-2188. Rudolph et al, p 1486.)* All 50 states in the United States, as well as most developed countries, screen in the neonatal period infants for a variety of conditions, among them galactosemia. The condition is autosomal recessive (about 1:40,000 live births); if not identified on newborn screening, affected infants can present with jaundice, hepatomegaly, vomiting, hypoglycemia, convulsions, lethargy, irritability, feeding problems, poor weight gain, aminoaciduria, cataracts, liver cirrhosis/failure, and mental retardation. Early treatment is essential and consists of galactose avoidance (soy or casein hydrolysate infant formula). With appropriate treatment, many of the features listed can be avoided. However, affected children often have ovarian failure, reduced bone mineral density, and developmental delay.

124. The answer is b. *(Hay et al, p 797. Kliegman et al, p 727. McMillan et al, p 425. Rudolph et al, pp 185-186.)* The clavicle is the most commonly fractured bone in the delivery process. While some fractures are identified at birth, others may not be identified until callus formation is noted at about a week of age. Clavicular fracture may happen in any delivery, although there is higher risk with large-for-gestational-age infants. Initial presentation of a fractured clavicle may include a pseudoparalysis, in which the infant refuses to move the ipsilateral arm, mimicking an Erb-Duchenne paralysis.

125. The answer is c. *(Hay et al, p 23. Kliegman et al, pp 720-721, 2826. McMillan et al, p 2492. Rudolph et al, p 2447.)* In a difficult delivery in which traction is applied to the head and neck, several injuries, including all those listed in the question, may occur. Erb-Duchenne paralysis affects the fifth and sixth cervical nerves; the affected arm cannot be abducted or externally rotated at the shoulder, and the forearm cannot be supinated. Injury to the seventh and eighth cervical and first thoracic nerves (Klumpke paralysis) results in palsy of the hand and also can produce Horner syndrome. Fractures in the upper limb are not associated with a characteristic posture, and passive movement usually elicits pain. Spinal injury causes complete paralysis below the level of injury. When paralysis of an upper extremity from injury to the brachial plexus is found in a neonate, injury to the phrenic nerve should also be suspected because the nerve roots are close together and can be injured concurrently. The paralyzed diaphragm can be noted to remain elevated on a chest x-ray taken during deep inspiration when it will contrast with the opposite normal diaphragm in its lower normal position; on expiration, this asymmetry cannot be seen. On inspiration, not only is breathing impaired since the paralyzed diaphragm does not contract, but also the negative pressure generated by the intact diaphragm pulls the mediastinum toward the normal side, impairing ventilation further. The diagnosis can easily be made by fluoroscopy, where these characteristic movements on inspiration and expiration can be seen. Rarely, both diaphragms can be paralyzed, producing much more severe ventilatory impairment. Fortunately, these injuries frequently improve spontaneously.

126. The answer is b. *(Hay et al, p 5. Kliegman et al, p 714. McMillan et al, p 197. Rudolph et al, pp 186-187.)* The child has a subgaleal or subaponeurotic hemorrhage which can be life threatening; infants may lose a third or more of their blood volume into this potential space, leading to hypovolemic shock. A subgaleal hemorrhage will feel like a cephalohematoma that crosses

the midline, but rapidly expands and can have cardiovascular complications. Careful monitoring is essential. Observation alone in this case would be appropriate, but should be accomplished in an ICU setting.

127. The answer is a. *(Hay et al, p 60. Kliegman et al, p 773. McMillan et al, p 445. Rudolph et al, pp 192-193.)* An infant of 2100 g (4 lb, 10 oz) at 38 weeks would be considered small for gestational age (SGA), a not uncommon consequence of maternal toxemia. Pregnancy-induced hypertension can produce a decrease in uteroplacental blood flow and areas of placental infarction. This can result in fetal nutritional deprivation and intermittent fetal hypoxemia, with a decrease in glycogen storage and a relative erythrocytosis, respectively. Hence, neonatal hypoglycemia and polycythemia are common clinical findings in these infants. A blood glucose level of 30 to 40 mg/dL in a full-term infant, however, is probably normal during the first postnatal day, and an infant is very unlikely to have a convulsion as a result of a level of 41 mg. Serum calcium levels usually decline during the first 2 to 3 postnatal days, but will only be considered abnormally low in a term infant when they fall below 7.5 to 8 mg/dL. Neonatal hypermagnesemia is common in an infant whose mother has received $MgSO_4$ therapy, but is usually asymptomatic or produces decreased muscle tone or floppiness. A persistent venous hematocrit of greater than 65% in a neonate is regarded as polycythemia and will be accompanied by an increase in blood viscosity.

Manifestations of the "hyperviscosity syndrome" include tremulousness or jitteriness that can progress to seizure activity because of sludging of blood in the cerebral microcirculation or frank thrombus formation, renal vein thrombosis, necrotizing enterocolitis, and tachypnea. Simple phlebotomy, while decreasing blood volume, will also decrease systemic arterial pressure and thus increase viscosity (based on Poiseuille law of flow). Therapy by partial exchange transfusion with saline or lactated Ringer solution is preferred, and may be more likely to be useful if performed prophylactically before significant symptoms have developed, but literature evaluating outcomes in these infants is lacking.

128. The answer is b. *(Hay et al, pp 29-30. Kliegman et al, pp 723-725. McMillan et al, pp 209-210. Rudolph et al, p 102.)* In the description provided, the most likely cause of the neonatal depression is maternal analgesic narcotic drug administration. While controlling the pain of the delivery in the mother, use of narcotics can result in depression of the newborn via crossing of the placenta. The appropriate first step in the management of this infant

(after managing the ABCs of airway, breathing, and circulation) is the administration of naloxone, 0.1 mg/kg, IM, IV, or endotracheal. The other possibilities are unlikely, given the clinical information provided.

129. The answer is a. *(Hay et al, p 30. Kliegman et al, pp 725-726. McMillan et al, p 211. Rudolph et al, p 196.)* Infants who are postmature (more than 42 weeks' gestation) and show evidence of chronic placental insufficiency (low birth weight for gestational age and wasted appearance) have a higher than average chance of being asphyxiated, and passage of meconium into the amniotic fluid thus places these infants at risk for meconium aspiration. Ideally the obstetrician suctions the mouth, nose, and hypopharynx immediately after delivery of the infant's head but before delivery of the reminder of the body. If the infant's heart rate is more than 100 beats per minute and respirations are unlabored, routine neonatal management is appropriate. However, if the heart rate is less than 100 beats per minute in a floppy, depressed infant then endotracheal intubation is accomplished along with suctioning and providing oxygen.

130. The answer is d. *(Hay et al, p 432. Kliegman et al, pp 2588-2589. McMillan et al, pp 811-812. Rudolph et al, p 2370.)* The time of onset of symptoms is somewhat helpful in the diagnosis of ophthalmia neonatorum. Chemical conjunctivitis is a self-limited condition that presents within 6 to 12 hours of birth and lasts for the first day or so of life; it is a consequence of ocular silver nitrate (no longer available in the United States) or erythromycin prophylaxis. Gonococcal conjunctivitis has its onset within 2 to 5 days after birth and is the most serious of the bacterial infections. Prompt and aggressive topical treatment and systemic antibiotics are indicated to prevent serious complications such as corneal ulceration, perforation, and resulting blindness. Parents should be treated to avoid the risk to the child of reinfection.

Chlamydial conjunctivitis occurs 5 to 14 days after birth; to avoid the risk of chlamydial pneumonia, treatment in an infant with conjunctivitis is with systemic antibiotics (parents, too, require treatment). However, asymptomatic infants born to chlamydia-positive mothers are not routinely treated with oral antibiotics at birth as prophylaxis, but rather watched closely for signs of infection, due to an increased incidence of hypertrophic pyloric stenosis among neonates having received erythromycin.

131. The answer is b. *(Hay et al, pp 35-36. Kliegman et al, pp 731-741. McMillan et al, pp 305-310. Rudolph et al, pp 127-134.)* For the child described

in the question, prematurity and the clinical picture presented make the diagnosis of hyaline membrane disease (infant respiratory distress syndrome) likely. HMD is caused by surfactant deficiency, and the incidence is increased with decreasing gestational age and birth weight. In this disease, lung compliance is reduced; lung volume is also reduced, and a significant right-to-left shunt of blood can occur. Some of the shunt can result from a patent ductus arteriosus or foramen ovale, and some can be due to shunting within the lung. Minute ventilation is higher than normal, and affected infants must work harder in order to sustain adequate respiration.

132. The answer is c. *(Hay et al, pp 1140-1151. Kliegman et al, pp 1435-1436. McMillan et al, pp 942-952. Rudolph et al, pp 1046, 1052.)* The transmission of HIV from mother to infant has decreased in recent years, due in large part to perinatal administration of antiretroviral medications to the mother and a course of zidovudine to the exposed infant. Studies suggest that a better than 50% decrease in transmission can be seen with appropriate medications as outlined.

An ELISA is an antibody test and will be positive in the infant born to an HIV-infected mother due to maternal antibodies that are passed through the placenta; it is not a useful test in the newborn infant to determine neonatal infection because of this expected transfer of maternal (and not infant) immunoglobulin. IVIG has not been shown to have a role in decreasing perinatal transmission. Healthy asymptomatic term infants born to HIV-infected mothers do not need special monitoring, nor do they need routine radiographs.

133. The answer is c. *(Hay et al, p 1032. Kliegman et al, pp 2386-2389. McMillan et al, pp 2635-2636. Rudolph et al, pp 2087-2089.)* Turner syndrome is a genetic disorder; the 45,XO karyotype is the most common. At birth, affected infants have low weights, short stature, edema over the dorsum of the hands and feet, and loose skin folds at the nape of the neck. Some other findings with this syndrome include sexual infantilism, streak gonads, typical faces, shield chest, low hairline, coarctation of the aorta, hypertension, bicuspid aortic valve, high palate, and horseshoe kidney. Coarse, tremulous movements accompanied by ankle clonus; vascular instability as evidenced, for example, by a harlequin color change (a transient, longitudinal division of a body into red and pale halves); softness of parietal bones at the vertex (craniotabes); and a liver that is palpable down to 2 cm below the costal margin are all findings often demonstrated by normal infants and are of no diagnostic significance in the clinical situation presented.

134. The answer is b. *(Hay et al, pp 271-273. Kliegman et al, pp 1682-1685. McMillan et al, pp 533-534. Rudolph et al, p 1498.)* The only vaccine routinely given in the newborn nursery is the hepatitis B vaccine. The other vaccines listed typically are administered beginning at 2 months of age. The other injection typically included on standardized order sheets and given to all hospitalized newborns is vitamin K, which is provided prophylactically to prevent a danger drop in vitamin K-dependent coagulation factors. Failure to provide vitamin K (especially those fed human breast milk) can result in vitamin K levels low enough to produce classic hemorrhagic manifestations (melena, hematuria, bleeding from circumcision site, intracranial hemorrhage, hypovolemic shock) on the second to seventh day of life.

135. The answer is b. *(Hay et al, p 404. Kliegman et al, p 2662. McMillan et al, p 828. Rudolph et al, p 1192.)* The pictured lesion is a Mongolian spot, a bluish-gray lesion located over the buttocks, lower back, and occasionally, the extensor surfaces of the extremities. These are common in blacks, Asians, and Latin Americans. They tend to disappear by 1 to 2 years of age, although those on the extremities may not fully resolve. Child abuse is unlikely to present with bruises alone; children frequently present with more extensive injuries. Subcutaneous fat necrosis, which may ultimately result in subcutaneous calcifications in the affected area, is usually found as a sharply demarcated, hard lesion on the cheeks, buttocks, and limbs but it usually is red. Hemophilia and vitamin K deficiency rarely present with subcutaneous lesions as described and are more likely to present as a bleeding episode.

136. The answer is c. *(Hay et al, p 609. Kliegman et al, pp 746-749. McMillan et al, pp 211-212. Rudolph et al, pp 189-191.)* Diaphragmatic hernia occurs with the transmittal of abdominal contents across a congenital or traumatic defect in the diaphragm. In the newborn, this condition results in profound respiratory distress with significant mortality. Prenatal diagnosis is common and, when found, necessitates that the birth take place at a tertiary-level center. In the neonate, respiratory failure in the first hours of life, a scaphoid abdomen, and presence of bowel sounds in the chest are common findings. Intensive respiratory support, including high-frequency oscillatory ventilation and extracorporeal membrane oxygenation (ECMO), has increased survival. Mortality can be as high as 50% despite aggressive treatment. While surgery may correct the diaphragmatic defect, the lung on the affected side remains hypoplastic and continues to contribute to morbidity.

Pneumonia and pneumothorax may cause respiratory distress with decreased breath sounds, but the radiograph in diaphragmatic hernia shows

the nasogastric tube curving into the left thorax, clearly an abnormal placement. A congenital cystic adenomatoid malformation will frequently look like a diaphragmatic hernia on radiographs, but the nasogastric tube would be in the correct location. Choanal atresia is an upper airway abnormality and does not cause these radiographic changes; it would have been difficult to place the nasogastric tube in the first place with this condition, in which there is a bony or membranous septum between the nose and pharynx.

137. The answer is d. *(Hay et al, p 46. Kliegman et al, pp 1543-1544. McMillan et al, pp 369-371. Rudolph et al, pp 1385-1387.)* Abdominal distention, choking, drooling, and coughing associated with feedings are symptoms of esophageal anomalies. The anomaly illustrated by **D** is the most common; that of **A** can be diagnosed after repeated episodes of pneumonia. The anomalies in **E** and **C** are associated with all the same symptoms except abdominal distention, which cannot develop because air cannot enter the gastrointestinal tract. **B** and **C** are the least common; in these, the upper esophageal segment is connected directly to the trachea, and massive entry of fluid into the lungs occurs.

VATER association, a complex of cardiovascular malformations, skeletal malformations, and renal abnormalities, has tracheoesophageal fistula as a common finding.

138. The answer is d. *(Hay et al, pp 271-273. Kliegman et al, pp 1682-1685. McMillan et al, pp 533-534. Rudolph et al, pp 49-50.)* The infant of a mother, who is a carrier of hepatitis B surface antigen, has a significant risk of acquiring infection. This usually occurs at the time of delivery, but infection can also be acquired during pregnancy and postnatally. A small percentage of infected neonates develop acute icteric hepatitis, but the majority remains asymptomatic. Of these infected asymptomatic infants, 80% or more will develop chronic infection, the long-term consequences of which are chronic liver disease and, possibly, hepatocellular carcinoma. Combined passive-active immunoprophylaxis in the form of hepatitis B immune globulin and hepatitis B vaccine affords protection not only from immediate perinatal infection but also from infection that may be acquired as a result of continued exposure in the household of a chronic carrier.

Immunization in this infant is indicated regardless of the presence of hepatitis B "e" antigen (HBeAg) in the mother. Although the presence of HBeAg, especially in the absence of antibody to HBeAg, is associated with high rates of transmission to neonates, any woman positive for hepatitis B surface antigen (HBsAg) is potentially infectious. It is not necessary to isolate

infants born to carriers of HBsAg, and screening of neonates for HBsAg is not indicated. Testing for HBsAg and anti-HBsAg at least 1 month after the third dose of hepatitis B vaccine will determine the efficacy of these measures.

139. The answer is a. *(Hay et al, p 25. Kliegman et al, p 693. McMillan et al, pp 171-173.)* The effect of a drug on the fetus is determined by the nature of the drug and by the timing and degree of exposure. Heparin does not cross the placental barrier and does not appear to directly affect the fetus once pregnancy is well established. Phenytoin may cause birth defects when given during the first trimester. Penicillin and aluminum hydroxide have not been found to affect the fetus. Propranolol, which may cause growth retardation when given throughout pregnancy, diminishes the ability of an asphyxiated infant to increase heart rate and cardiac output. It has also been associated with hypoglycemia and apnea.

140. The answer is b. *(Hay et al, pp 12-15. Kliegman et al, pp 758-759. McMillan et al, pp 240-241. Rudolph et al, pp 164-169.)* The development of jaundice in a healthy full-term baby may be considered the result of a normal physiologic process in certain circumstances. It may be normal if the time of onset and duration of the jaundice and the pattern of serially determined serum concentrations of bilirubin are in conformity with currently accepted safe criteria. Physiologic jaundice becomes apparent on the second or third day of life, peaks to levels no higher than about 12 mg/dL on the fourth or fifth day, and disappears by the end of the week. The rate of rise is less than 5 mg/dL per 24 hours and levels of conjugated bilirubin do not exceed about 1 mg/dL. Concern about neonatal jaundice relates to the risk of the neurotoxic effects of unconjugated bilirubin.

The precise level and duration of exposure necessary to produce toxic effects are not known, but bilirubin encephalopathy, or kernicterus, is rare in term infants whose bilirubin level is kept below 18 to 20 mg/dL. Certain risk factors affecting premature or sick newborns increase their susceptibility to kernicterus at much lower levels of bilirubin. The diagnosis of physiologic jaundice is made by excluding other causes of hyperbilirubinemia by means of history, physical examination, and laboratory determinations.

Jaundice appearing in the first 24 hours is usually a feature of hemolytic states and is accompanied by an indirect hyperbilirubinemia, reticulocytosis, and evidence of red-cell destruction on smear. In the absence of blood group or Rh incompatibility, congenital hemolytic states (eg, spherocytic anemia) or G6PD deficiency should be considered. With infection, hemolytic and hepatotoxic factors are reflected in the increased levels of both direct and indirect

bilirubin. Studies should include maternal and infant Rh types and blood groups and Coombs tests to detect blood-group or Rh incompatibility and sensitization.

Measurements of total and direct bilirubin concentrations help to determine the level of production of bilirubin and the presence of conjugated hyperbilirubinemia. Hematocrit and reticulocyte count provide information about the degree of hemolysis and anemia, and a complete blood count screens for the possibility of sepsis and the need for cultures.

Examination of the blood smear is useful in differentiating common hemolytic disorders. Except for determinations of total and direct bilirubin, tests of liver function are not particularly helpful in establishing the cause of early-onset jaundice. Transient elevations of transaminases (AST and ALT) related to the trauma of delivery and to hypoxia have been noted.

Biliary atresia and neonatal hepatitis can be accompanied by elevated levels of transaminase, but characteristically present as chronic cholestatic jaundice with mixed hyperbilirubinemia after the first week of life.

141. The answer is d. *(Hay et al, p 47. Kliegman et al, pp 1559-1560. McMillan et al, p 373. Rudolph et al, pp 1403-1404.)* The finding of polyhydramnios suggests high intestinal obstruction, signs of which include abdominal distention and early and repeated regurgitation. Distention usually is not present, as vomiting keeps the intestine decompressed. The bile-stained vomitus of the infant places the obstruction distal to the ampulla of Vater, eliminating esophageal atresia and pyloric stenosis from consideration. The "double bubble" sign on the x-ray is characteristic of duodenal atresia, which is compatible with the history. Midgut volvulus, which may obstruct the bowel in the area of the duodenojejunal junction, most often produces signs after an affected infant is 3 or 4 days old with acute onset of bilious vomitus. Gastric duplication does not usually produce intestinal obstruction; a cystic mass may be palpated on abdominal examination. Patients with duodenal atresia should be examined closely for evidence of other conditions such as Down syndrome or heart disease.

142. The answer is b. *(Kliegman et al, pp 939-940, 1611-1612. McMillan et al, pp 1448-1449. Rudolph et al, p 1332.)* Goat's milk, by itself, is not an ideal source of infant nutrition as it contains inadequate folate and iron, potentially contributing to anemia. Caloric content is actually more dense than cow's milk–based formulas (about 30 kcal/oz, compared to 20 kcal/oz for formula and breast milk). Casein and whey are the protein sources in goat's milk, as

they are for many formulas. Some manufacturers in different countries produce a goat milk–based formula that contains supplemental vitamins and iron. Unpasteurized goat's milk should never be used as an infant formula because of the risk of brucellosis.

143. The answer is d. *(Hay et al, pp 450-451. Kliegman et al, pp 1532-1533. McMillan et al, pp 469-472, 2661-2662. Rudolph et al, pp 748-753.)* The infant pictured has bilateral cleft lip and palate. This defect occurs in about 4% of the siblings of affected infants; its incidence in the general population is 1 in 1000. Evaluation for other structural and chromosomal abnormalities is indicated. Although affected infants are likely to have feeding problems initially, these problems usually can be overcome by feeding in a propped-up position and using special nipples. Complications include recurrent otitis media and hearing loss as well as speech defects, which may be present despite good anatomic closure. Repair of a cleft lip usually is performed within the first 2 to 3 months of life; the palate is repaired later, usually between the ages of 6 months and 5 years.

144. The answer is a. *(Hay et al, pp 947-950. Kliegman et al, pp 2319-2325. McMillan et al, pp 421-422, 2126-2128. Rudolph et al, pp 2065-2070.)* The clinical findings of congenital hypothyroidism are subtle, and may not be present at all at birth; this is thought to be a result of passage of some T_4 transplacentally. Infants with examination findings will usually have an umbilical hernia and a distended abdomen. The head may be large, and the fontanelles will be large as well. The child may be hypothermic and have feeding difficulties; constipation and jaundice may be persistent. Skin may be cold and mottled, and edema may be found in the genitals and extremities. The heart rate may be slow, and anemia may develop. As these findings may be subtle or nonexistent, neonatal screening programs are extremely important for early diagnosis of these infants.

Sepsis can cause hypothermia and poor feeding, but the 2-week time makes this choice unlikely. Hirschsprung disease may cause chronic constipation and abdominal distension, but not the other findings. Botulism can cause a flaccid paralysis and poor feeding, but the large fontanelles and umbilical hernia are not caused by this infection.

145. The answer is d. *(Hay et al, pp 1024, 1044. Kliegman et al, pp 697, 2444-2447. McMillan et al, pp 175, 265-266. Rudolph et al, pp 2186-2187.)* Diseases that are due to defects in a single gene are designated as autosomal or X-linked,

depending on whether the affected gene is located on an autosome or an X chromosome. Genetically determined diseases that are multifactorial in origin (ie, neural tube defects) do not conform to the Mendelian pattern of inheritance but exhibit a variable outcome that reflects the interaction between a particular genotype and an environment. The relatives of persons with diseases of multifactorial origin have an increased risk of having similar abnormalities.

The recurrence risk for most single primary defects of multifactorial inheritance (eg, neural tube defects) is increased with each child affected. This increased risk forms the basis for assuming that genetic factors play a role in the occurrence of these abnormalities. Other factors, such as race, sex, and ethnic and racial background, influence the frequency with which an abnormality of multifactorial inheritance occurs in relatives. The prenatal diagnosis of neural tube defects (anencephaly and meningomyelocele) can be made by the detection of elevated levels of alpha-fetoprotein in the amniotic fluid. To reduce the risk of neural tube defects, it is now recommended that all women capable of becoming pregnant take 400 mcg of folic acid daily.

146. The answer is e. *(Hay et al, p 20. Kliegman et al, p 741. McMillan et al, p 311. Rudolph et al, pp 179-181.)* Transient tachypnea of the newborn is usually seen after a normal vaginal or, especially, after a cesarean delivery. The condition is a result of retained fetal lung fluid. These patients have tachypnea, retractions, grunting, and sometimes cyanosis. The chest examination is usually normal; the chest radiograph demonstrates prominent pulmonary vascular markings with fluid in the fissures and hyperexpansion (flat diaphragms). Therapy is supportive, with maintenance of normal oxygen saturation. Resolution usually occurs in the first 3 days of life.

Diaphragmatic hernia would demonstrate intestinal contents located in the chest on plain radiograph. Meconium aspiration would present in a patient who is born with meconium at the delivery and the radiograph would have patchy infiltrates. Pneumonia in a patient of this age is more likely to have diffuse infiltrates on radiograph and clinical findings of temperature instability. Idiopathic respiratory distress syndrome occurs in premature infants whose radiographs demonstrate a "ground glass appearance."

147. The answer is d. *(Hay et al, pp 1205-1208. Kliegman et al, pp 1263-1269. McMillan et al, pp 1135-1142. Rudolph et al, pp 1004-1005.)* The clinical presentation of congenital syphilis is varied. Many newborns appear normal at birth and continue to be asymptomatic for the first few weeks or months of life. Most untreated infants will develop skin lesions, the usual one being an

infiltrative, maculopapular peeling rash that is most prominent on the face, palms, and soles. Involvement of the nasal mucous membranes causes rhinitis with a resultant serous, and occasionally purulent, blood-tinged discharge (snuffles). This, as well as scrapings from the skin lesions, contains abundant viable treponemes. Hepatosplenomegaly and lymphadenopathy are common, and early jaundice is a manifestation of syphilitic hepatitis. Liver function tests are elevated; hemolytic anemia and thrombocytopenia are common. Infants may have a saddle nose, a result of destruction of bone from syphilitic rhinitis. Among the later manifestations, or stigmata, of congenital syphilis is interstitial keratitis, which is an acute inflammation of the cornea that begins in early childhood (most commonly between 6 and 14 years of age). Interstitial keratitis represents the response of the tissue to earlier sensitization. Findings include marked photophobia, lacrimation, corneal haziness, and eventual scarring. Hutchinson teeth (peg or barrel-shaped upper central incisors), abnormal enamel, and mulberry molars (first lower molars with an abnormal number of cusps) are dental manifestations of syphilis.

148. The answer is b. *(Kliegman et al, pp 2829-2930. McMillan et al, p 2123. Rudolph et al, p 2493.)* Fifth-finger polydactyly is 10 times more common in black than in white children and is typically familial. This finding in otherwise healthy black children should raise no special concern. In a white child, careful examination of the cardiac system is warranted. Similarly, preaxial (or thumb-side) polydactyly is unusual and should be further investigated. Some syndromes that are associated with polydactyly are trisomy 13, Rubinstein-Taybi syndrome, Meckel-Gruber syndrome, and Ellis-van Creveld syndrome. Simple postaxial polydactyly without bone involvement may be surgically removed shortly after birth. Simply tying these appendages off with string will usually leave a nub.

149. The answer is c. *(Kliegman et al, p 693. Rudolph et al, p 2196.)* Infants born to narcotic addicts are more likely than other children to exhibit a variety of problems, including perinatal complications, prematurity, and low birth weight. The onset of withdrawal commonly occurs during an infant's first 2 days of life and is characterized by hyperirritability and coarse tremors, along with vomiting, diarrhea, fever, high-pitched cry, and hyperventilation; seizures and respiratory depression are less common. The production of surfactant can be accelerated in the infant of a heroin-addicted mother.

In utero exposure to alcohol leads to a fetal alcohol syndrome consisting of growth retardation, microcephaly, flat philtrum, thin upper lip, cardiac

defects, and hypoplastic fifth fingernails. Maternal cocaine use puts the infant at risk for vascular accidents and premature delivery, but not a withdrawal syndrome. Similarly, marijuana is not associated with neonatal withdrawal. Maternal tobacco use can result in small-for-gestation age infants, but not a neonatal withdrawal syndrome as described in the question.

150. The answer is b. *(Kliegman et al, p 711. McMillan et al, p 346. Rudolph et al, pp 1934-1936.)* Idiopathic apnea is common in premature infants but is not expected in the full-term newborn. When apnea occurs in the term infant, there is almost always an identifiable cause. Sepsis, gastroesophageal reflux, congenital heart disease, seizures, hypoglycemia, and airway obstruction can cause apnea in term newborns. Harlequin syndrome is a transient change in the skin color of the otherwise asymptomatic newborn (usually preterm) in which the dependent side of the entire body turns red while the upper side remains pale.

151. The answer is d. *(Hay et al, pp 49-53, 763, 1088-1090. Kliegman et al, pp 2515-2516. McMillan et al, pp 493-496. Rudolph et al, pp 106-107.)* Neonatal sepsis, a clinical syndrome of systemic illness accompanied by bacteremia, often results in spread of infection to the meninges and other distant sites. The diagnosis of serious infection, including meningitis, in a neonate is difficult because the signs and symptoms are subtle and nonspecific. They include lethargy; feeding problems including abdominal distention, vomiting, and diarrhea; temperature instability; respiratory distress or apnea; and jaundice. Nuchal rigidity and Kernig and Brudzinski signs are frequently not present in the neonate with meningitis.

152. The answer is b. *(Hay et al, pp 25-26. Kliegman et al, pp 699-701. Rudolph et al, pp 63, 68, 118-119.)* Twin-to-twin transfusions occur in about 15% of monochorionic twins and commonly cause intrauterine death. This disorder should be suspected when the hematocrits of twins differ by more than 15 mg/dL. The donor twin is likely to have oligohydramnios, anemia, and hypovolemia with evidence of shock if the hematocrit is significantly reduced; the recipient twin is likely to have hydramnios and plethora and to be larger than the donor twin. A 20% difference in body weight may result. As the central venous hematocrit rises above 65%, infants can develop hyperviscosity, respiratory distress, hyperbilirubinemia, hypocalcemia, renal vein thrombosis, congestive heart failure, and convulsions.

153. The answer is b. *(Hay et al, pp 32-34. Kliegman et al, p 681. McMillan et al, pp 200-201. Rudolph et al, pp 82, 99, 105, 168, 207.)* There is loss of

body weight of 1.5% to 2% per day for the first 5 days of life for a normal newborn infant as excessive fluid is excreted. One might think this would tend to cause hemoconcentration and produce an increase in hematocrit, but, to the contrary, the hematocrit falls as an adaptation to an environment of higher oxygen. Infants usually have several meconium stools during the first day or two of life, changing to soft yellow stools after 1 to 2 days of life. As the hematocrit falls, there is a corresponding increase in serum bilirubin that peaks around 3 to 5 days of life. Temperature should not change; temperature instability in a term infant is frequently a sign of serious infection.

154. The answer is c. *(Hay et al, pp 46, 1044. Kliegman et al, pp 685-686. McMillan et al, pp 191-192, 373 1850. Rudolph et al, pp 78, 191, 1639.)* It is generally presumed that duodenal atresia and tracheoesophageal fistula lead to hydramnios (polyhydramnios) by interference with reabsorption of swallowed amniotic fluid. Hydramnios is also associated with approximately 80% of infants who have trisomy 18. Approximately 50% of women with anencephalic fetuses have polyhydramnios. Oligohydramnios occurs in association with congenital abnormalities of the fetal kidneys or other parts of the genitourinary tract, such as renal agenesis or obstruction, that impede normal formation or excretion of fetal urine.

155. The answer is b. *(Hay et al, pp 49-58. Kliegman et al, pp 794-811. McMillan et al, pp 482-492. Rudolph et al, pp 86, 106-107.)* Although hypothyroid neonates may develop hyperbilirubinemia, the patient described most likely has a congenital or acquired infection requiring immediate diagnosis and, if possible, treatment. Among the important causes of neonatal sepsis are prenatal infections, including congenital syphilis, toxoplasmosis, cytomegalic inclusion disease, and rubella. Useful diagnostic studies, in addition to cultures for bacteria, include specific serologic tests for pathogens, viral cultures, lumbar puncture, and x-rays of the chest and long bones. Longitudinal striations in the metaphyses are characteristic of congenital rubella, whereas osteochondritis or periostitis usually indicates congenital syphilis. Congenital syphilis, cytomegalovirus, and rubella can be highly contagious. Urine can contain rubella virus for more than 6 months and is, therefore, a special hazard to nonimmune pregnant women.

156-159. The answers are 156-c, 157-e, 158-a, 159-d. *(Hay et al, pp 53-55. Kliegman et al, pp 1340, 1361, 1377-1379, 1486-1496. McMillan et al, pp 511-520, 528-532. Rudolph et al, pp 169-173, 1033, 1077-1079, 1147.)*

Congenital rubella infection affects all organ systems. Infants will be small, with intrauterine growth retardation. They may also manifest cataracts, microphthalmia, myocarditis, and a red or purple macular rash ("blueberry muffin" rash). Structural heart defects (such as a patent ductus arteriosis, pulmonary artery stenosis, and septal defects) are typical of congenital rubella, but not in the other TORCH infections. Laboratory anomalies may include a hemolytic anemia with thrombocytopenia, elevated liver functions, and pleocytosis in the spinal fluid. Affected children do not have a good prognosis. Congenital rubella is not commonly seen in developed countries with high immunization rates.

Transmission of HSV from mother to newborn can happen *in utero*, intrapartum, and postnatally. Intrapartum transmission is most common. Infants born vaginally to a mother with a primary genital herpes infection are at highest risk for disease, with up to 50% risk of perinatal transmission. About half the infants of congenital HSV are born to mothers who are unaware of their infection. Infants can display isolated CNS involvement, isolated cutaneous infection, or systemic generalized infection. Treatment usually is with acyclovir; even with therapy, morbidity is high in infants with CNS involvement.

Toxoplasmosis can cause symptoms similar to other congenital infections, but the combination of hydrocephalus, chorioretinitis, and intracranial calcifications are considered the "classic triad" of toxoplasma infection in a neonate. Infection usually occurs during primary infection of the mother or as a reactivation of infection in an immune-compromised host. These infants may also display symptoms similar to other congenital infections, such as anemia, a petechial rash, organomegaly, jaundice, and seizures.

Congenital cytomegalovirus is the most common sort of congenital infection, with infection estimates ranging from 0.4% to 2.4% of all live births. Many cases are asymptomatic; others may develop cytomegalic inclusion disease, a multiorgan manifestation of disease including intrauterine growth restriction (IUGR), hepatosplenomegaly, jaundice, petechiae or purpura, microcephaly, chorioretinitis, and intracranial calcifications. More than half of infants with this congenital infection develop sensorineural hearing loss.

160-163. The answers are 160-i, 161-f, 162-e, 163-d. *(Hay, pp 23, 506-507, 536, 1106-1108, 1044. Kliegman et al, pp 721, 726, 750-752, 1773-1777, 1783-1784. McMillan et al, pp 212, 313-314, 699-700, 1391-1394, 2492. Rudolph et al, pp 89, 148-151, 186, 191, 1984-1985.)* Infants with upper

brachial plexus injury (cervical nerves 3, 4, 5) can also have ipsilateral phrenic nerve paralysis. These infants can present with labored, irregular breathings and cyanosis; the injury is usually unilateral. Confirmation of the diagnosis is made with ultrasound or fluoroscopy which confirms "seesaw" movements during respiration of the two sides of the diaphragm.

Pneumothorax occurs with a frequency of about 1% to 2% of births, but they are rarely symptomatic. Incidence is higher in infants born with meconium-stained fluid, and the chest radiograph is as that described. Transillumination may assist in the diagnosis while awaiting radiograph; immediate treatment for infants with significant distress is with a 23-guage butterfly needle attached to a stopcock and removal of the air. For those without significant distress and who are not on high levels of ambient oxygen, 100% oxygen therapy can assist in nitrogen washout.

Primary pulmonary hypoplasia (Potter sequence) includes a dysmorphic child (widely spaced eyes, low set ears, broad nose, receding chin, limb abnormalities) and bilateral renal agenesis, which leads to oligohydramnios. These infants have immediate respiratory distress; the condition is not compatible with life.

Bronchiolitis is a very common viral infection most often caused by respiratory syncytial virus. It is most often seen in the winter months with symptoms of wheezing, hypoxia, and respiratory distress seen in younger children; often an older sibling has milder, upper respiratory symptoms. Premature infants, infants with congenital heart disease, infants with a variety of lung disorders, and infants with immune system defects are at higher risk of severe complications. Diagnosis is made by clinical history and/or detection of the viral antigen in nasal secretions; treatment is supportive.

164-167. The answers are 164-a, 165-a, 166-c, 167-b. *(Hay et al, pp 996-997. McMillan et al, pp 2126-2127, 2153-2154, 2171-2172, 2187-2188. Rudolph et al, pp 609-611, 626, 640-641, 1486, 2065-2070.)* In galactosemia, an enzyme deficiency (galactose-l-phosphate uridyl transferase) results in a block in the metabolic pathway of galactose and leads to the accumulation of galactose-l-phosphate in the tissues. Infants with this condition develop serious damage to liver, brain, and eyes after being fed milk containing lactose (a disaccharide compound of glucose and galactose). Clinical manifestations include lethargy, vomiting and diarrhea, hypotonia, hepatomegaly and jaundice, failure to thrive, and cataracts. The course of the disease in untreated patients is variable; death from liver failure and inanition can occur; most untreated patients develop physical and mental retardation. Treatment consists

of prompt elimination of lactose-containing milk from the diet in infancy and, as a more varied diet is introduced, exclusion of foods that contain casein, dry milk solids, whey, or curds.

Phenylketonuria, a genetically determined disorder with an autosomal recessive pattern of inheritance, is caused by the absence of an enzyme that metabolizes phenylalanine to tyrosine. The resultant accumulation of phenylalanine and its metabolites in the blood leads to severe mental retardation in untreated patients. Treatment consists of a diet that maintains phenylalanine at levels low enough to prevent brain damage but adequate to support normal physical and mental development. Careful supervision of the low-phenylalanine diet and monitoring of blood levels are necessary. Special formulas are available for the infant; older children have difficulty following the diet. It is not clear when and if the diet can be discontinued.

Biotinidase is the enzyme responsible for breakdown of biocytin (the lysyl precursor of biotin) to free biotin. Deficiency of the enzyme, which is inherited as an autosomal recessive trait, results in malfunctioning of the biotin-dependent mitochondrial enzymes and in organic acidemia. Clinical problems related to the deficiency appear several months or years after birth and include dermatitis, alopecia, ataxia, hypotonia, seizures, developmental delay, deafness, immunodeficiency, and metabolic acidosis. The treatment is lifelong administration of free biotin.

The treatment of congenital hypothyroidism with oral levothyroxine sodium should begin as early as possible to prevent psychomotor retardation. Periodic measurement of T3, T4, and TSH is necessary to assess the response to therapy and the need for adjustment of the dose of thyroxine. Careful evaluation of somatic growth by plotting sequential measurements and monitoring bone age is essential.

168-171. The answers are 168-c, 169-a, 170-b, 171-b. *(Hay et al, pp 435, 474, 750-751, 903-905. Kliegman et al, pp 2140-2143, 2484-2487 2621. McMillan et al, pp 1775-1777, 2385-2387, 2659-2660. Rudolph et al, pp 770-772, 1185, 1614-1616, 2347-2348.)* Aniridia is found in 1% to 2% of children with Wilms tumor. Genitourinary anomalies are found in 4% to 5%, and hemihypertrophy is associated with this tumor in 2% to 3% of patients. Wilms tumor is the most common primary renal malignancy in childhood. Presentation is usually an abdominal mass, sometimes with hypertension, hematuria, abdominal pain, and fever. Prognosis is generally good.

Waardenburg syndrome is inherited as an autosomal dominant trait with variable penetrance. It includes, in decreasing order of frequency, the

following anomalies: lateral displacement of the medial canthi, broad nasal bridge, medial hyperplasia of the eyebrows, partial albinism commonly expressed by a white forelock or heterochromia (or both), and deafness in 20% of cases.

A flat capillary vascular malformation in the distribution of the trigeminal nerve is the basic lesion in the Sturge-Weber syndrome. The malformation also involves the meninges and results in atrophy to the underlying cerebral cortex. The damage is manifested clinically by grand mal seizures, mental deficiency, and hemiparesis or hemianopsia on the contralateral side. The cause is unknown.

Infants who have tuberous sclerosis are often born with hypopigmented oval or irregularly shaped skin macules (ash leaf). Cerebral sclerotic tubers also present from birth and become visible radiographically by the third to fourth year of life. Myoclonic seizures, present in infancy, can convert to grand mal seizures later in childhood. Adenoma sebaceum appears at 4 to 7 years of age. The disease, which also affects the eyes, kidneys, heart, bones, and lungs, is inherited as an autosomal dominant trait with variable expression; new mutations are very common.

172-174. The answers are 172-e, 173-c, 174-a. *(Hay et al, pp 5, 23, 39-41, 63, 428. Kliegman et al, pp 171-178, 405, 714-717. McMillan et al, pp 150-152, 197, 271-276. Rudolph et al, pp 144, 186-187, 2242.)* A subgaleal or subaponeurotic hemorrhage can be life threatening; infants may lose a third or more of their blood volume into this potential space, leading to hypovolemic shock. A subgaleal hemorrhage will feel like a cephalohematoma that crosses the midline. Careful monitoring is essential.

Subdural hematomas are commonly seen as part of the shaken baby syndrome. This lesion occurs when the bridging cortical veins that drain the cerebral cortex have been ruptured, leading to a collection of blood between the dura and the cerebral mantle. Repeated trauma can lead to additional collections of blood. In many children, additional findings of abuse such as broken bones, bruises, and retinal hemorrhages are found.

Caput succedaneum is soft-tissue swelling of the scalp involving the presenting delivery portion of the head. This lesion is sometimes ecchymotic and can extend across the suture lines. The edema resolves in the first few days of life.

Intraventricular hemorrhage (IVH) is commonly seen in very small, preterm infants. The incidence of IVH increases with smaller-size infants and in those with perinatal complications. It occurs in the gelatinous

subependymal germinal matrix of the brain and can lead to progressive posthemorrhagic hydrocephalus. Hydrocephalus in these children can present with enlarging head circumference, apnea and bradycardia, lethargy, bulging fontanelle, widely split sutures, or no signs at all. Therapy can include ventricular-peritoneal shunting.

Cephalohematomas do not cross the suture line, since they are subperiosteal hemorrhages. No discoloration of the scalp is seen, and the swelling usually progresses over the first few hours of life. Occasionally, skull fractures are present as well. Most cephalhematomas resolve within the first few weeks or months of life without residual findings.